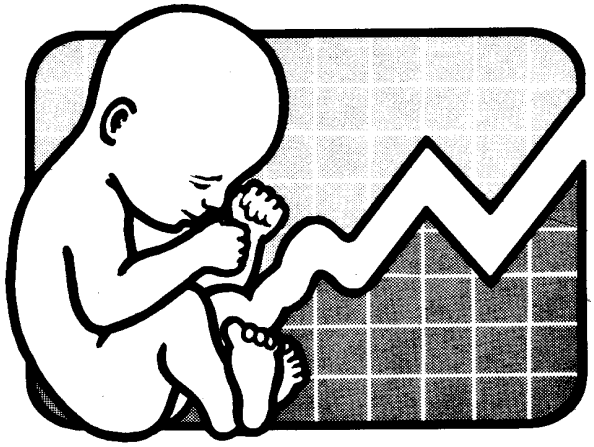
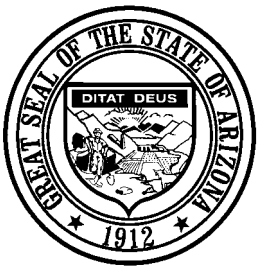
Arizona Department of Health Services

1994 and 1997 Arizona Birth Defects Monitoring Program Report





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1994 and 1997
ARIZONA BIRTH DEFECTS MONITORING PROGRAM REPORT

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EXECUTIVE SUMMARY

In 1994, there were 1,315 infants with a reportable birth defect among 70,896 live births and 507 stillbirths in Arizona. In 1997, there were 1,401 infants with reportable birth defects among 75,563 live births and 637 stillbirths. This report presents the rates for 44 composite categories of birth defects (encompassing 140 conditions) developed by the Centers for Disease Control and Prevention (CDC). These categories represent the most serious defects. Arizona's overall birth defect rate in 1994 was 18.4 for live born and stillborn cases per 1,000 live births and stillbirths combined. In 1997 the overall rate was also 18.4 cases per 1,000 births, which was not significantly different from the 1996 birth defect rate of 18.6 cases per 1,000 births. Pyloric stenosis (152 cases), oral clefts (146 cases), obstruction of kidney/ureter (110 cases), Down Syndrome (101 cases), microcephaly (69 cases), and hip dislocation (55 cases) were the most common birth defects in 1997, as they were in 1994.

Race/Ethnicity Patterns

In 1997, the state rate for spina bifida (including children with and without hydrocephalus) was slightly higher than the state rate for 1996, but the rates have remained relatively constant since 1989. Although Hispanics had lower overall rates of spina bifida than Whites did in 1993 and 1994, they have had higher rates than Whites since 1995 (as they did before 1993). Hispanics and Whites had the highest rates for microcephaly in 1994, followed by Native Americans. In 1997, Native Americans and Blacks had the highest rates for microcephaly, followed by Hispanics and Whites. In both years the pyloric stenosis rates were highest for Whites, followed by Native Americans. Rates for both racial groups were higher than the overall state rates for pyloric stenosis in 1994 and 1997. Blacks and Native Americans exhibited Down Syndrome rates higher than the state rates in 1997. Cleft lip with and without cleft palate rates for Hispanics and Native Americans were higher than the state rate in 1994. By 1997, however, Native Americans had the highest rate for cleft lip with and without cleft palate–a rate significantly higher ($p < .01$) than the state rate that year. The second highest rates for clefts in 1997 were among Whites. Hispanics were third highest. For gastroschisis, Hispanics had a higher rate than Whites in both years. Fetal alcohol syndrome rates (FAS) were significantly higher ($p < .01$) among Native Americans than they were for the state as a whole in both years.

Age Patterns

In 1994 and in 1997 infants born to women older than 34 were significantly more likely to have

birth defects than were babies born to Arizona women as a whole ($p < .01$). In 1997, significantly more infants born to teens had birth defects that than those who were born to the general population (at $p < .01$). The incidence of Down Syndrome (Trisomy 21) increased with maternal age and was significantly higher ($p < .01$) for mothers who were over 34 years in 1994 and 1997. The incidence of Down Syndrome was significantly lower ($p < .01$) for women 25 to 29 years old in 1997. In contrast, higher gastroschisis rates were associated with younger maternal age in both years.

County Patterns

The number of live-born infants with a reportable birth defect by county are aggregated for the years 1986 through 1997 to provide numbers large enough for analysis. Gila county had the highest rate of infants with congenital anomalies, followed by Navajo, Graham and Pinal counties. In contrast, Greenlee and La Paz counties had the lowest rates. Nevertheless, tests of significance show that there are no statistical differences between the overall birth defect rate of the counties and the state rate.

THE IMPORTANCE OF ARIZONA'S BIRTH DEFECTS REGISTRY

The Arizona Birth Defects Monitoring Program (ABDMP) is a population-based registry which provides information on the occurrence rates for birth defects. The registry provides for ongoing surveillance to monitor trends and detect possible problems early.[1,2,3] The information is used to direct and evaluate birth defect prevention efforts and to direct allocation of resources for health services. Such a registry is necessary because other systems for reporting birth defects, including birth certificates and hospital discharge data, are not accurate or complete due to under-reporting of cases, lack of specificity of birth defects, and incomplete recording of birth defect information.[4]

To ensure that it is able to provide accurate and current information for health planning and prevention activities, the ABDMP has entered into a three-year Cooperative Agreement with the Centers of Disease Control and Prevention, starting in the fall of 2000, to set up a rapid birth defect surveillance system (The ABDMP Rapid Surveillance System). This surveillance system will identify infants born in Arizona with anencephaly, encephalocele, spina bifida, cleft lip or cleft palate within three months of birth, and will provide affected families with information and resources for follow-up medical and social services.

The system is currently being established in Maricopa and Pima Counties and will be set up statewide in 2003. Possible cases of these birth defects will be identified in prenatal testing facilities; hospital logs and medical records; clinics; birth, death and fetal death certificates; the Newborn Screening Program and through Children's Rehabilitative Services. Medical records will be reviewed promptly to verify diagnoses prior to entering the cases into the birth defects registry database. Once cases have been matched with birth and fetal death certificates, parents of living children with these defects will be contacted by letter and given a packet of general information about their child's defect, a fact sheet summarizing state and community resources relevant to their child's diagnosis, application forms for state programs which provide follow-up services, and recurrence prevention information. A follow-up phone call will be made to families whose children are not enrolled in Children's Rehabilitative Services at six months of age and community resource referral and recurrence prevention information will be reviewed. In this way the ABDMP will hopefully increase the percentage of children with these severe birth defects receiving coordinated, multi-disciplinary follow-up services to minimize the long-term disabilities associated with these birth defects. It is hoped that this program will also increase the number of families of children with birth defects receiving support from other families of

children with similar difficulties. Furthermore, this rapid-surveillance system will enable the Arizona Department of Health Services Folate Committee to track trends in neural tube defect occurrence and evaluate the efficacy of programs educating the public on folic acid for neural tube defect prevention.

Economic Cost

Birth defects remain the leading cause of infant mortality in both Arizona and the United States. Arizona data show that between 1989 and 2000, one out of four infant deaths was due to a birth defect. In contrast, infant deaths attributable to low birth weight and due to SIDS have decreased, accounting only for less then one out of every ten infant deaths (see Figure 1).[5, 6] Birth defects also remain the fifth leading cause of years of potential life lost. With the advancement in technology, many infants with birth defects survive beyond their first birthday, often requiring special medical services, education and rehabilitation services, vocational training and in some cases, custodial care, incurring a lifetime of costs and hardships for their families.

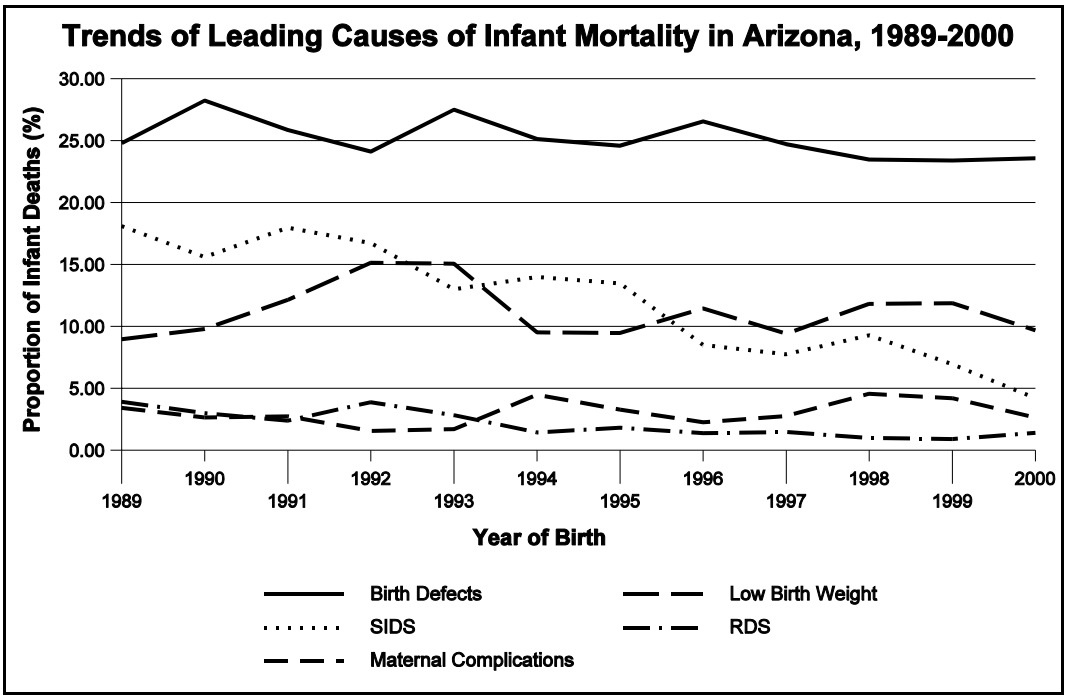


Figure 1. Trends of Leading Causes of Infant mortality in Arizona, 1989-2000

Birth Defect Surveillance in the United States

Like diseases, injuries, and other chronic conditions, birth defects are monitored conditions, similar to infections. Birth defect surveillance programs were first established in response to clusters of birth defects which were associated with the mother's use of medication such as thalidomide or concerns over environmental pollutants. Lately, birth defect surveillance programs are increasingly used to implement and evaluate intervention and prevention programs. Information from these programs can also be used to facilitate research efforts to be able to identify the etiology of birth defects[7].

Three major categories of known causes of birth defects are chromosomal errors, environmental factors, and maternal illness, infections or conditions during pregnancy. For example, maternal alcohol consumption may cause fetal alcohol syndrome and German measles in early pregnancy may cause congenital rubella syndrome. In spite of research efforts in identifying the etiology of birth defects, causes of most birth defects remain unknown. In recent years, maternal intake of the B-vitamin folic acid has been shown to be necessary for proper fetal development. Among some populations the consumption of adequate folic acid (400 micrograms daily) prior to conception and through the first trimester has reduced the incidence of neural tube defects (NTDs) by at least 50%.[8] There is also research that shows that folic acid may reduce the incidence of certain types of heart defects, urinary tract defects and oral clefts.[9,10,11] A considerable amount of research still needs to be undertaken to augment our knowledge on birth defects, its etiology and its impact on different groups. If Arizona is to ensure the well-being of its children, it is essential that the surveillance and documentation of the occurrence of birth defects in the state be undertaken and made available to the public and researchers.

METHODS

The ABDMP is a statewide, population-based, active ascertainment program, pursuant to Arizona Revised Statute §36-133, which mandates the surveillance of chronic diseases, including birth defects. The funding for the ABDMP comes from appropriations of the state legislature to the Arizona Department of Health Services. Trained ABDMP staff collect data from 64 reporting sources including 58 hospitals; two centers providing genetics services; four state Children's Rehabilitative Services clinics; and the state Office of Vital Records. Ascertainment procedures used by the ABDMP are nearly identical to those used by the California Birth Defects Monitoring Program and the U.S. Centers for Disease Control's Metropolitan Atlanta Congenital Defects Program (MACDP).

Hospital case-finding sources include the disease index, labor and delivery log, nursery log, newborn intensive care log, pediatric log, and pathology/autopsy log. Potential cases are also identified through a review of the Hospital Discharge Data Base, and Birth, Death and Fetal Death Certificates. All records for children up to one year of age with recorded birth defects become part of this listing of possible cases. The medical records of possible cases are reviewed to determine which records meet the case definition. An abstract (case report) of the medical record is completed for each reportable case. In order to identify children born with birth defects in 1997, ABDMP staff reviewed 5,923 medical records, identified reportable cases, and excluded those not meeting the case definition. The abstracts of cases identified from multiple sources are compared, merged, and added to the registry.

If the nature of a defect diagnosed in the first year of life is more precisely diagnosed later in the child's life and this information is contained in the chart at the time of our review (which may occur several years after the child's birth or fetal death) then the more precise diagnosis is used. (See Appendix 3.)

ABDMP staff assign a six-digit classification code to each defect. The classification system is CDC's modification of the British Pediatric Association (BPA) Classification of Disease. This coding system is similar to the International Classification of Disease (ICD). The staff collect diagnostic information on birth defects that fall primarily within the range of ICD-9-CM Codes 740.00-759.99. The system of codes is hierarchical: the more digits in the code, the more precise the diagnosis. ABDMP staff always code the data at the most precise level possible.

CASE DEFINITION

The following are the criteria for inclusion in the Birth Defects Monitoring Program case file:

A. The mother's place of residence at the time of birth must be in Arizona. (Charts of mothers or babies residing in Mexico are not abstracted).

B. The child must have a structural, genetic, or biochemical birth defect, or other specified birth outcome that can adversely affect the infant's health and development. (Most, but not all, are listed in ICD-9-CM 740.0-759.9.)

C. The defect must be diagnosed, or signs and symptoms of a potential defect recognized, within the first year of life.

D. Stillborn infants are included if they have a reportable birth defect.

E. The date of birth (or delivery for stillbirths > 19 weeks of gestational age) is on or after January 1, 1986.

F. For a list of exclusions, see Appendix 5.

Due to the need to collect and report data on birth defects in a more timely manner, effective March 1996, the ABDMP reduced the number of reportable conditions from over 500 to 140 conditions. The 140 conditions include only the major congenital anomalies recommended by The International Clearinghouse for Birth Defects Monitoring Systems and recommended by CDC. The retained, reportable defects still permit the ABDMP to compare its rates with other registries for the major birth defects categories. The reduced list of reportable defects applies to data collected on births after 1991. Since this change in 1996, data have been collected much quicker for births in 1992 through 1997. At present data are being collected for infants born from 1998 to 2000.

INTERPRETING THE DATA

The tables and figures presented in this report represent data collected on birth defects in Arizona for the period 1986 to 1997. Each table presents the reported counts, rates and confidence intervals on selected congenital anomalies. Below is an explanation of how counts, rates, and confidence intervals were calculated.

Counts

The counts, sometimes called cases, represent the number of children who were diagnosed with a reportable birth defect within the first year of life. Children born with more than one reportable defect, as often occurs, may be part of the counts across the 44 composite categories.

Rates

Incidence rates of birth defects were calculated by dividing the number of children (cases) with a particular reportable defect by the total number of live births (and in some cases live births plus fetal deaths) for the specific year of interest, and then multiplying by 10,000 or 1,000. In most tables and figures we show rates that are calculated by including live births and fetal deaths in both the numerator and denominator. For example, there were 101 children (live born and stillborn infants >= 20 weeks of gestation) with Down Syndrome in 1997. There were 76,200 births (live births plus fetal deaths) in 1997. The rate is calculated as such: (101/76,200)*10,000 = 13.25 cases of Down Syndrome per 10,000 live births and fetal deaths.

Confidence Intervals

The confidence intervals shown in the tables and figures are provided to give information about the estimate of the rate. Confidence intervals presented in this report are 99 percent Poisson confidence intervals. The confidence intervals indicate that the true rate should be contained in this interval 99 percent of the time. For example, Down Syndrome in 1997 occurs at a rate of 13.25 per 10,000 births. The lower and upper bounds of the point estimate of this rate are 10.1 and 17.1, respectively. Thus, one can say that 99 percent of the time the true rate of Down Syndrome is between 10.1 and 17.1 cases per 10,000 live births and fetal deaths.

Small Numbers and a Note Of Caution

While the intent of these data is to provide the reader with useful information on birth defects in Arizona, it is equally important not to mislead data users. Rates, confidence intervals, and any other analyses based on fewer than 10 reported cases cannot be considered statistically reliable and are not shown for local areas.

Tests of Significance

Z tests were used to determine whether there were statistically significant differences in the rates between groups and areas. The state rate was used as the standard rate in these tests.

STATE PROFILE OF DEFECTS

The Arizona Birth Defects Monitoring Program (ABDMP) has been in operation since 1986 and has collected data since that time. This is the eleventh annual report of data compiled by the ABDMP in its mission to collect and analyze information on children with birth defects and to provide data for the study of the causes of birth defects in Arizona.

Tables and Figures

Table 1-A presents data on the 44 categories of birth defects collected by the ABDMP among live born and stillborn infants by race for 1994. Table 1-B presents similar data for 1997. Tables 2-A and 2-B display the number of live born and stillborn infants with all reportable birth defects (140 conditions) and the average number of defects among live born and stillborn infants by county for 1994 and 1997, respectively. Fetal deaths include therapeutic abortions and stillborn babies with reportable congenital defects if the estimated gestational age was greater than 19 weeks. Table 3 displays rates of the 44 categories of congenital anomalies by year for 1986 through 1997. The series of graphs in Figure 2 display the trends for the same categories of defects.

Race/Ethnicity

A section on the rates of selected birth defects by race/ethnicity follows the section on the state profile. Included in this section are spina bifida with and without hydrocephaly, abdominal wall defects, Down Syndrome, microcephaly, pyloric stenosis, and fetal alcohol syndrome.

Maternal Age

Figures 10 to 12 focus on the relationship of birth defects and maternal age. Figure 10 presents birth defect rates by maternal age groupings and the confidence interval of these rates for each of the age groups. Figures 11 and 12 display the rates for Down Syndrome and gastroschisis by maternal age groupings.

County Profiles

The last section of the report presents aggregated data on the number of live-born infants with a reportable (as modified in 1996) birth defect between 1986 to 1997 by county and by race. Aggregated rates for selected birth defects among live-born infants by county are also shown.

Table 1-A
Arizona Birth Defects Monitoring Program
Congenital Anomalies by Race/Ethnicity - Arizona 1994
Incidence Rates[a,b] per 10,000 Live Births and Fetal Deaths

CODE	DEFECT GROUP (Composite Category)	TOTAL	RATE	WHITE NON-HISP.	RATE	HISP.	RATE	BLACK	RATE	NATIVE AMER.	RATE	OTHER	RATE
A00	CENTRAL NERVOUS SYSTEM												
A01	Anencephaly	28	3.92	13	3.42	11	4.61	1	3.98	3	5.39	0	0.00
A02	Spina bifida with hydrocephaly	19	2.66	11	2.89	7	2.93	0	0.00	1	1.80	0	0.00
A03	Spina bifida without hydrocephaly	16	2.24	11	2.89	2	0.84	0	0.00	3	5.39	0	0.00
A13	Encephalocele	11	1.54	3	0.79	7	2.93	1	3.98	0	0.00	0	0.00
A15	Hydrocephaly	34	4.76	17	4.47	13	5.44	2	7.96	2	3.59	0	0.00
A16	Microcephaly	75	10.50	26	6.83	26	10.89	5	19.91	17	30.54	1	7.09
B00	EYE AND EAR												
B03	Glaucoma	4	0.56	2	0.53	1	0.42	0	0.00	1	1.80	0	0.00
B04	Cataract	14	1.96	5	1.31	6	2.51	0	0.00	3	5.39	0	0.00
B51	Anophthalmia	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
B52	Microphthalmia	18	2.52	6	1.58	10	4.19	2	7.96	0	0.00	0	0.00
B54	Ear anomaly with hearing loss	33	4.62	11	2.89	12	5.03	2	7.96	8	14.37	0	0.00
D00	CARDIAC												
D01	Truncus arteriosus	6	0.84	3	0.79	1	0.42	1	3.98	1	1.80	0	0.00
D02	Transposition of great vessels	30	4.20	19	4.99	7	2.93	3	11.95	1	1.80	0	0.00
D03	Tetralogy of Fallot	30	4.20	15	3.94	11	4.61	0	0.00	4	7.19	0	0.00
D04	Single ventricle	5	0.70	3	0.79	2	0.84	0	0.00	0	0.00	0	0.00
D51	Aortic stenosis	21	2.94	13	3.42	6	2.51	1	3.98	0	0.00	1	7.09
D52	Hypoplastic left heart	9	1.26	6	1.58	2	0.84	1	3.98	0	0.00	0	0.00
D53	Total anomalous pulmonary venous return	6	0.84	3	0.79	2	0.84	0	0.00	1	1.80	0	0.00
E00	RESPIRATORY												
E01	Choanal atresia	10	1.40	8	2.10	1	0.42	1	3.98	0	0.00	0	0.00
E06	Agenesis of lung	19	2.66	11	2.89	6	2.51	0	0.00	2	3.59	0	0.00
F00	OROFACIAL AND GASTROINTESTINAL												
F01	Cleft palate	45	6.30	23	6.05	15	6.28	1	3.98	5	8.98	1	7.09
F02	Cleft lip with and without cleft palate	83	11.62	43	11.30	31	12.98	2	7.96	7	12.58	0	0.00
F08	Pyloric stenosis	159	22.27	103	27.08	39	16.33	2	7.96	13	23.36	2	14.17
F09	Tracheoesophageal fistula	14	1.96	6	1.58	7	2.93	0	0.00	1	1.80	0	0.00

[a] Incidence rates include live born and stillborn cases in numerator and denominator.
[b] Incidence rates based on counts of less than 10 events are not statistically reliable.

Table 1-A Continued
Arizona Birth Defects Monitoring Program
Congenital Anomalies by Race/Ethnicity- Arizona 1994
Incidence Rates[a,b] per 10,000 Live Births and Fetal Deaths

CODE	DEFECT GROUP (Composite Category)	TOTAL	RATE	WHITE NON-HISP.	RATE	HISP.	RATE	BLACK	RATE	NATIVE AMER.	RATE	OTHER	RATE
F00	OROFACIAL AND GASTROINTESTINAL												
F14	Stenosis/atresia of duodenum	14	1.96	5	1.31	6	2.51	1	3.98	2	3.59	0	0.00
F15	Stenosis/atresia of small intestine	15	2.10	5	1.31	6	2.51	0	0.00	4	7.19	0	0.00
F16	Stenosis/atresia of rectum	25	3.50	14	3.68	10	4.19	1	3.98	0	0.00	0	0.00
F17	Hirschsprung's disease	10	1.40	4	1.05	6	2.51	0	0.00	0	0.00	0	0.00
F18	Malrotation of intestine	24	3.36	13	3.42	6	2.51	3	11.95	1	1.80	1	7.09
F21	Biliary atresia	6	0.84	4	1.05	2	0.84	0	0.00	0	0.00	0	0.00
H00	GENITO-URINARY												
H01	Renal agenesis	25	3.50	11	2.89	11	4.61	1	3.98	2	3.59	0	0.00
H06	Obstruction of kidney/ureter	86	12.04	46	12.09	32	13.40	3	11.95	5	8.98	0	0.00
H09	Bladder or urethra obstruction	11	1.54	7	1.84	4	1.68	0	0.00	0	0.00	0	0.00
J00	MUSCULOSKELETAL												
J03	Dislocation of hip	65	9.10	29	7.62	24	10.05	0	0.00	11	19.76	1	7.09
J51	Complete absence of upper or lower limb	1	0.14	1	0.26	0	0.00	0	0.00	0	0.00	0	0.00
J52	Phocomelia of limb	2	0.28	1	0.26	1	0.42	0	0.00	0	0.00	0	0.00
K05	Amniotic bands	9	1.26	3	0.79	4	1.68	0	0.00	1	1.80	1	7.09
N01	Diaphragmatic hernia	21	2.94	12	3.15	5	2.09	0	0.00	2	3.59	2	14.17
N02	Omphalocele	11	1.54	4	1.05	4	1.67	1	3.98	2	3.59	0	0.00
N04	Gastroschisis	27	3.78	11	2.89	14	5.86	0	0.00	2	3.59	0	0.00
R00	SYNDROMES												
R01	Down Syndrome (Trisomy 21)	100	14.00	43	11.30	45	18.85	3	11.95	9	16.17	0	0.00
R02	Patau Syndrome (Trisomy 13)	3	0.42	1	0.26	2	0.84	0	0.00	0	0.00	0	0.00
R03	Edwards Syndrome (Trisomy 18)	14	1.96	10	2.63	2	0.84	0	0.00	2	3.59	0	0.00
S02	Fetal alcohol syndrome	14	1.96	1	0.26	0	0.00	1	3.98	12	21.56	0	0.00

[a] Incidence rates include live born and stillborn cases in numerator and denominator.
[b] Incidence rates based on counts of less than 10 events are not statistically reliable.

Table 1-B
Arizona Birth Defects Monitoring Program
Congenital Anomalies by Race/Ethnicity - Arizona 1997
Incidence Rates[a,b] per 10,000 Live Births and Fetal Deaths

CODE	DEFECT GROUP (Composite Category)	TOTAL	RATE	WHITE NON-HISP.	RATE	HISP.	RATE	BLACK	RATE	NATIVE AMER.	RATE	OTHER	RATE
A00	CENTRAL NERVOUS SYSTEM												
A01	Anencephaly	24	3.15	9	2.36	13	4.62	0	0.00	1	1.88	1	4.99
A02	Spina bifida w/ hydrocephaly	24	3.15	12	3.14	10	3.55	0	0.00	2	3.76	0	0.00
A03	Spina bifida w/o hydrocephaly	14	1.83	6	1.57	8	2.84	0	0.00	0	0.00	0	0.00
A13	Encephalocele	9	1.18	1	0.26	6	2.13	1	3.98	1	1.88	0	0.00
A15	Hydrocephaly	31	4.07	18	4.71	10	3.55	0	0.00	3	5.64	0	0.00
A16	Microcephaly	69	9.06	23	6.02	32	11.36	4	15.91	9	16.92	1	4.99
B00	EYE AND EAR												
B03	Glaucoma	3	0.40	0	0.00	2	0.71	1	3.98	0	0.00	0	0.00
B04	Cataract	13	1.71	6	1.57	4	1.42	2	7.96	1	1.88	0	0.00
B51	Anophthalmia	1	0.13	1	0.26	0	0.00	0	0.00	0	0.00	0	0.00
B52	Microphthalmia	18	2.36	8	2.09	4	1.42	1	3.98	5	9.40	0	0.00
B54	Ear Anomaly w/ hearing loss	47	6.17	18	4.71	16	5.68	0	0	11	20.68	2	9.98
D00	CARDIAC												
D01	Truncus Arteriosus	11	1.44	3	0.78	6	2.13	1	3.98	0	0.00	1	4.99
D02	Transposition of great vessels	40	5.25	17	4.45	17	6.04	2	7.96	3	5.64	1	4.99
D03	Tetralogy of Fallot	33	4.33	16	4.19	8	2.84	3	11.93	5	9.40	1	4.99
D04	Single ventricle	11	1.44	7	1.83	3	1.06	0	0.00	1	1.88	0	0.00
D51	Aortic stenosis	27	3.54	13	3.40	13	4.62	0	0.00	1	1.88	0	0.00
D52	Hypoplastic left heart	12	1.57	8	2.09	4	1.42	0	0.00	0	0.00	0	0.00
D53	Total anomalous pulmonary venous return	8	1.05	3	0.78	3	1.06	0	0.00	2	3.76	0	0.00
E00	RESPIRATORY												
E01	Choanal atresia	11	1.44	8	2.09	1	0.36	2	7.96	0	0.00	0	0.00
E06	Agenesis of lung	18	2.36	5	1.31	8	2.84	0	0.00	5	9.40	0	0.00
F00	OROFACIAL AND GASTROINTESTINAL												
F01	Cleft palate	43	5.64	19	4.97	17	6.04	2	7.96	4	7.52	1	4.99
F02	Cleft lip w/ & w/o cleft palate	103	13.52	49	12.83	29	10.30	1	3.98	22	41.35	2	9.98
F08	Pyloric stenosis	152	19.95	88	23.04	45	15.98	5	19.89	12	22.56	2	9.98
F09	Tracheoesophageal fistula	21	2.75	11	2.88	7	2.49	3	11.93	0	0.00	0	0.00

[a] Incidence rates include live born and stillborn cases in numerator and denominator.
[b] Incidence rates based on counts of less than 10 events are not statistically reliable.

Table 1-B Continued
Arizona Birth Defects Monitoring Program
Congenital Anomalies by Race/Ethnicity - Arizona 1997
Incidence Rates[a,b] per 10,000 Live Births and Fetal Deaths

CODE	DEFECT GROUP (Composite Category)	TOTAL	RATE	WHITE NON-HISP.	RATE	HISP.	RATE	BLACK	RATE	NATIVE AMER.	RATE	OTHER	RATE
F00	OROFACIAL AND GASTROINTESTINAL												
F14	Stenosis/atresia of duodenum	11	1.44	4	1.05	6	2.13	1	3.98	0	0.00	0	0.00
F15	Stenosis/atresia of small intestine	12	1.57	6	1.57	5	1.78	1	3.98	0	0.00	0	0.00
F16	Stenosis/atresia of rectum	16	2.10	8	2.09	7	2.49	1	3.98	0	0.00	0	0.00
F17	Hirschsprung's disease	8	1.05	6	1.57	2	0.71	0	0.00	0	0.00	0	0.00
F18	Malrotation of intestine	16	2.10	6	1.57	7	2.49	3	11.93	0	0.00	0	0.00
F21	Biliary atresia	2	0.26	2	0.52	0	0.00	0	0.00	0	0.00	0	0.00
H00	GENITO-URINARY												
H01	Renal agenesis	34	4.46	15	3.93	13	4.62	1	3.98	2	3.76	3	14.96
H06	Obstruction of kidney or ureter	110	14.44	47	12.30	49	17.40	2	7.96	9	16.92	3	14.96
H09	Bladder or urethra obstruction	10	1.31	5	1.31	5	1.78	0	0.00	0	0.00	0	0.00
J00	MUSCULOSKELETAL												
J03	Dislocation of hip	55	7.22	25	6.54	21	7.46	1	3.98	8	15.04	0	0.00
J51	Complete absence of upper or lower limb	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
J52	Phocomelia of limb	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
K05	Amniotic bands	12	1.57	6	1.57	5	1.78	0	0.00	1	1.88	0	0.00
N01	Diaphragmatic hernia	15	1.97	7	1.83	6	2.13	0	0.00	1	1.88	1	4.99
N02	Omphalocele	11	1.44	6	1.57	4	1.42	0	0.00	1	1.88	0	0.00
N04	Gastroschisis	36	4.72	9	2.36	20	7.10	5	19.89	2	3.76	0	0.00
R00	SYNDROMES												
R01	Down Syndrome (Trisomy 21)	101	13.25	44	11.52	38	13.49	6	23.87	12	22.56	1	4.99
R02	Patau Syndrome (Trisomy 13)	5	6.56	1	0.26	3	1.07	0	0.00	0	0.00	1	4.99
R03	Edwards Syndrome (Trisomy 18)	15	1.97	7	1.83	5	1.78	1	3.98	2	3.76	0	0.00
S02	Fetal Alcohol Syndrome	9	1.18	2	0.52	1	0.36	0	0.00	6	11.28	0	0.00

[a] Incidence rates include live born and stillborn cases in numerator and denominator.
[b] Incidence rates based on counts of less than 10 events are not statistically reliable.

Table 2-A
Arizona Birth Defects Monitoring Program [a, b]
Birth Defects by County of Residence, 1994
(For the 140 Birth Defect Categories Monitored)

STATE, COUNTY	LIVE BORNS (LB) WITH DEFECTS		STILLBORNS (SB) WITH DEFECTS		LIVE BORNS AND STILLBORNS WITH DEFECTS		NUMBER OF DEFECTS IN LIVE-BORN INFANTS		NUMBER OF DEFECTS IN STILLBORN INFANTS	
	Number	% OF ALL LB W/ DEFECTS	Number	% OF ALL SB W/ DEFECTS	Number	% OF BIRTHS W/ DEFECTS	Number	AVG # PER CHILD	Number	AVG # PER CHILD
ARIZONA	1265	1.78	50	9.86	1315	1.84	2032	1.61	82	1.64
APACHE COUNTY	26	1.77	2	12.5	28	1.89	47	1.81	3	1.50
COCHISE COUNTY	26	1.53	1	12.5	27	1.58	50	1.92	1	1.00
COCONINO COUNTY	26	1.50	0	0.00	26	1.49	45	1.73	0	0.00
GILA COUNTY	10	1.49	0	0.00	10	1.47	13	1.30	0	0.00
GRAHAM COUNTY	9	2.16	0	0.00	9	2.14	13	1.44	0	0.00
GREENLEE COUNTY	3	2.00	0	0.00	3	2.00	3	1.00	0	0.00
LA PAZ COUNTY	3	1.35	0	0.00	3	1.34	3	1.00	0	0.00
MARICOPA COUNTY	773	1.83	27	8.77	800	1.89	1197	1.55	46	1.70
MOHAVE COUNTY	27	1.45	2	13.33	29	1.54	40	1.48	2	1.00
NAVAJO COUNTY	45	2.51	1	7.69	46	2.55	81	1.80	2	2.00
PIMA COUNTY	204	1.79	8	12.12	212	1.85	348	1.71	10	1.25
PINAL COUNTY	42	2.06	1	5.26	43	2.09	74	1.76	1	1.00
SANTA CRUZ COUNTY	10	1.30	0	0.00	10	1.30	21	2.10	0	0.00
YAVAPAI COUNTY	14	1.02	3	17.65	17	1.22	25	1.79	7	2.33
YUMA COUNTY	47	1.56	5	25.00	52	1.72	72	1.53	10	2.00

[a]Total number of live births in Arizona for 1994 = 70,896. [b]Total number of fetal deaths in Arizona for1994 = 507

Table 2-B
Arizona Birth Defects Monitoring Program [a, b]
Birth Defects by County of Residence, 1997
(For the 140 Birth Defect Categories Monitored)

STATE, COUNTY	LIVE BORNS (LB) WITH DEFECTS		STILLBORNS (SB) WITH DEFECTS		LIVE BORNS AND STILLBORNS WITH DEFECTS		NUMBER OF DEFECTS IN LIVE-BORN INFANTS		NUMBER OF DEFECTS IN STILLBORN INFANTS	
	Number	% OF ALL LB W/ DEFECTS	Number	% OF ALL SB W/ DEFECTS	Number	% OF BIRTHS W/ DEFECTS	Number	AVG # PER CHILD	Number	AVG # PER CHILD
ARIZONA	1334	1.77	67	10.52	1401	1.84	2213	1.66	124	1.85
APACHE COUNTY	29	2.18	2	15.38	31	2.31	52	1.79	5	2.50
COCHISE COUNTY	21	1.27	2	20.00	23	1.39	35	1.67	3	1.50
COCONINO COUNTY	25	1.39	1	5.00	26	1.43	57	2.28	1	1.00
GILA COUNTY	16	2.41	1	12.50	17	2.50	38	2.37	3	3.00
GRAHAM COUNTY	9	1.83	1	33.3	10	2.00	13	1.44	1	1.00
GREENLEE COUNTY	1	0.60	0	0.00	1	0.59	1	1.00	0	0.00
LA PAZ COUNTY	3	1.50	0	0.00	3	1.49	8	2.67	0	0.00
MARICOPA COUNTY	818	1.74	44	11.40	862	1.81	1326	1.62	78	1.77
MOHAVE COUNTY	30	1.70	1	16.67	31	1.75	54	1.80	1	1.00
NAVAJO COUNTY	33	1.99	1	6.67	34	2.03	74	2.24	1	1.00
PIMA COUNTY	227	2.00	12	12.12	239	2.08	351	1.55	28	2.33
PINAL COUNTY	38	1.77	0	0.00	38	1.74	76	2.00	0	0.00
SANTA CRUZ COUNTY	15	1.93	0	0.00	15	1.92	19	1.27	0	0.00
YAVAPAI COUNTY	24	1.55	1	5.55	25	1.60	41	1.71	1	1.00
YUMA COUNTY	45	1.57	1	4.35	46	1.59	68	1.51	2	2.00

[a]Total number of live births in Arizona for 1997 = 75,563. [b]Total number of fetal deaths in Arizona for 1997 = 637.

Table 3
Congenital Anomalies by Year, 1986 - 1997
Incidence Rates[a] Per 1,000 Live Births and Fetal Deaths, Arizona

CODE/CONDITION*		1986	1987	1988	1989	1990	1991	1992	1993	1994	1995	1996	1997
A01 A nencephaly	Cases	22	17	18	18	16	17	21	15	28	18	22	24
	Rate	0.35	0.26	0.27	0.27	0.23	0.25	0.30	0.22	0.39	0.25	0.29	0.31
	CI	0.19-0.60	0.12-0.48	0.13-0.48	0.13-0.48	0.11-0.43	0.12-0.45	0.16-0.52	0.10-0.41	0.23-0.63	0.12-0.44	0.16-0.49	0.17-0.52
A02 Spina bifida with hydrocephaly	Cases	26	24	19	22	23	21	26	21	19	24	20	24
	Rate	0.42	0.37	0.28	0.33	0.33	0.31	0.38	0.30	0.26	0.33	0.26	0.31
	CI	0.24-0.69	0.20-0.62	0.14-0.50	0.17-0.55	0.18-0.56	0.16-0.53	0.21-0.61	0.16-0.52	0.13-0.47	0.18-0.55	0.14-0.46	0.17-0.52
A03 Spina bifida without hydrocephaly	Cases	10	11	6	16	16	13	12	14	16	10	13	14
	Rate	0.16	0.17	0.09	0.24	0.23	0.19	0.17	0.20	0.22	0.14	0.17	0.18
	CI	0.06-0.35	0.06-0.35	0.02-0.23	0.11-0.44	0.11-0.43	0.08-0.37	0.07-0.35	0.09-0.39	0.11-0.41	0.05-0.29	0.07-0.34	0.08-0.35
A13 Encephalocele	Cases	10	8	14	5	13	14	2	6	11	7	16	9
	Rate	0.16	0.12	0.21	0.07	0.19	0.20	0.03	0.09	0.15	0.10	0.21	0.12
	CI	0.06-0.35	0.03-0.29	0.09-0.40	0.02-0.21	0.08-0.37	0.09-0.39	0.00-0.13	0.02-0.23	0.06-0.32	0.03-0.24	0.10-0.39	0.04-0.26
A15 Hydrocephaly	Cases	34	41	48	44	52	46	34	28	34	40	35	31
	Rate	0.55	0.64	0.72	0.65	0.75	0.67	0.49	0.40	0.48	0.55	0.46	0.41
	CI	0.34-0.85	0.41-0.95	0.48-1.04	0.43-0.95	0.51-1.06	0.44-0.97	0.30-0.75	0.23-0.64	0.29-0.73	0.35-0.82	0.29-0.71	0.24-0.64
A16 Microcephaly	Cases	30	60	70	109	118	120	90	83	75	81	67	69
	Rate	0.49	0.94	1.06	1.61	1.70	1.75	1.30	1.19	1.05	1.11	0.89	0.91
	CI	0.29-0.77	0.65-1.30	0.76-1.43	1.17-1.96	1.33-2.15	1.37-2.21	0.97-1.70	0.88-1.57	0.76-1.41	0.82-1.47	0.63-1.21	0.65-1.23
B03 Glaucoma	Cases	2	7	4	5	4	2	1	3	4	5	5	3
	Rate	0.03	0.10	0.06	0.07	0.06	0.03	0.01	0.04	0.06	0.07	0.07	0.04
	CI	0.04-0.15	0.03-0.26	0.00-0.19	0.02-0.21	0.01-0.18	0.00-0.14	0.00-0.11	0.00-0.16	0.01-0.18	0.01-0.19	0.01-0.19	0.01-0.17

* See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.
“Cases” are the number of live born and stillborn infants $20 weeks gestation.

Table 3 Continued
Congenital Anomalies by Year, 1986 - 1997
Incidence Rates[a] Per 1,000 Live Births and Fetal Deaths, Arizona

CODE/CONDITION*		1986	1987	1988	1989	1990	1991	1992	1993	1994	1995	1996	1997
B04 Cataract	Cases	8	7	7	15	24	10	12	8	14	14	9	13
	Rate	0.13	0.10	0.10	0.22	0.35	0.15	0.17	0.11	0.20	0.19	0.12	0.17
	CI	0.04-0.30	0.03-0.26	0.03-0.26	0.10-0.42	0.19-0.57	0.05-0.31	0.07-0.35	0.04-0.27	0.09-.038	0.09-0.37	0.04-0.27	0.07-0.34
B51 Anophthalmia	Cases	6	1	3	5	7	5	3	2	0	2	1	1
	Rate	0.09	0.01	0.04	0.07	0.10	0.07	0.04	0.03	0.00	0.03	0.01	0.01
	CI	0.02-0.25	0.00-0.11	0.00-0.16	0.02-0.21	0.03-0.25	0.02-0.21	0.00-0.16	0.00-0.13	0.00-0.00	0.00-0.13	0.00-0.10	0.00-0.10
B52 Microphthalmia	Cases	10	24	21	19	24	29	22	14	18	24	19	18
	Rate	0.16	0.37	0.31	0.28	0.35	0.42	0.32	0.20	0.25	0.33	0.25	0.24
	CI	0.06-0.35	0.20-0.62	0.16-0.54	0.14-0.50	0.19-0.57	0.25-0.67	0.17-0.54	0.09-0.39	0.12-0.45	0.18-0.55	0.13-0.44	0.12-0.42
B54 Hearing loss with ear anomaly	Cases	33	59	34	50	59	65	41	42	33	44	34	47
	Rate	0.53	0.92	0.51	0.74	0.85	0.95	0.59	0.60	0.46	0.60	0.45	0.62
	CI	0.32-0.83	0.64-1.28	0.31-0.79	0.50-1.06	0.59-1.18	0.67-1.30	0.38-0.88	0.39-0.89	0.28-0.71	0.39-0.88	0.28-0.69	0.41-0.89
D01 Truncus arteriosus	Cases	4	10	9	9	6	6	3	4	6	3	7	11
	Rate	0.06	0.15	0.13	0.13	0.09	0.09	0.04	0.06	0.08	0.04	0.09	0.14
	CI	0.01-0.20	0.05-0.33	0.04-0.30	0.05-0.30	0.02-0.23	0.02-0.23	0.00-0.16	0.01-0.18	0.02-0.22	0.00-0.15	0.03-0.23	0.06-0.30
D02 Transposition of great vessels	Cases	32	26	26	33	28	26	25	28	30	33	34	40
	Rate	0.52	0.40	0.39	0.49	0.40	0.38	0.36	0.40	0.42	0.45	0.45	0.52
	CI	0.31-0.81	0.23-0.66	0.22-0.64	0.30-0.75	0.23-0.65	0.21-0.62	0.20-0.59	0.23-0.64	0.25-0.66	0.28-0.70	0.28-0.69	0.34-0.78
D03 Tetralogy of Fallot	Cases	15	18	29	23	27	22	32	30	30	29	34	33
	Rate	0.24	0.28	0.43	0.34	0.39	0.32	0.46	0.43	0.42	0.40	0.45	0.43
	CI	0.11-0.46	0.13-0.50	0.25-0.69	0.19-0.57	0.22-0.63	0.17-0.54	0.28-0.72	0.25-0.68	0.25-0.66	0.23-0.63	0.28-0.69	0.26-0.67

* See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.
“Cases” are the number of live born and stillborn infants $20 weeks gestation.

Table 3 Continued
Congenital Anomalies by Year, 1986 - 1997
Incidence Rates[a] Per 1,000 Live Births and Fetal Deaths, Arizona

CODE/CONDITION*													
		1986	1987	1988	1989	1990	1991	1992	1993	1994	1995	1996	1997
D04 Single ventricle	Cases Rate CI	2 0.03 0.00-0.15	4 0.06 0.01-0.19	5 0.07 0.01-0.21	4 0.06 0.01-0.19	6 0.09 0.02-0.23	1 0.01 0.00-0.11	3 0.04 0.00-0.16	8 0.11 0.04-0.27	5 0.07 0.01-0.20	5 0.07 0.01-0.19	9 0.12 0.04-0.27	11 0.14 0.06-0.30
D51 Aortic stenosis	Cases Rate CI	8 0.13 0.04-0.30	15 0.23 0.10-0.44	17 0.25 0.12-0.46	25 0.37 0.21-0.61	17 0.25 0.12-0.45	17 0.25 0.12-0.45	23 0.32 0.18-0.56	15 0.22 0.10-0.41	21 0.29 0.15-0.50	30 0.41 0.24-0.65	21 0.28 0.15-0.48	27 0.35 0.20-0.57
D52 Hypoplastic left heart	Cases Rate CI	9 0.14 0.05-0.32	16 0.25 0.11-0.46	8 0.12 0.03-0.28	16 0.24 0.11-0.44	19 0.28 0.14-0.48	11 0.16 0.06-0.33	13 0.19 0.08-0.37	14 0.20 0.09-0.39	9 0.13 0.04-0.28	10 0.14 0.05-0.29	15 0.20 0.09-0.37	12 0.16 0.06-0.32
D53 Total anomalous pulmonary venous return	Cases Rate CI	5 0.08 0.17-0.23	5 0.07 0.01-0.22	13 0.19 0.08-0.38	17 0.25 0.12-0.46	13 0.19 0.08-0.37	11 0.16 0.06-0.33	11 0.16 0.06-0.33	11 0.16 0.06-0.33	6 0.08 0.02-0.22	12 0.16 0.07-0.33	13 0.17 0.07-0.34	8 0.10 0.03-0.24
E01 Choanal atresia	Cases Rate CI	6 0.09 0.24-0.25	10 0.15 0.05-0.33	10 0.15 0.05-0.32	16 0.24 0.11-0.44	6 0.09 0.02-0.23	5 0.07 0.02-0.21	6 0.09 0.02-0.23	7 0.10 0.03-0.25	10 0.14 0.05-0.30	14 0.19 0.09-0.37	18 0.24 0.12-0.43	11 0.14 0.06-0.30
E06 Agenesis of lung	Cases Rate CI	25 0.40 0.22-0.67	44 0.69 0.45-1.00	32 0.48 0.29-0.75	42 0.62 0.40-0.92	49 0.71 0.47-1.01	50 0.73 0.49-1.04	26 0.38 0.21-0.61	21 0.30 0.16-0.52	19 0.27 0.13-0.47	14 0.19 0.09-0.37	23 0.30 0.17-0.51	18 0.24 0.12-0.42
F01 Cleft palate	Cases Rate CI	39 0.63 0.40-0.95	46 0.72 0.47-1.04	36 0.54 0.33-0.82	43 0.64 0.41-0.93	38 0.55 0.35-0.82	31 0.45 0.27-0.71	27 0.39 0.22-0.63	49 0.70 0.47-1.01	45 0.63 0.41-0.92	47 0.64 0.43-0.93	41 054 0.35-0.80	43 0.56 0.37-0.83

* See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.
“Cases” are the number of live born and stillborn infants $20 weeks gestation.

Table 3 Continued
Congenital Anomalies by Year, 1986 - 1997
Incidence Rates[a] Per 1,000 Live Births and Fetal Deaths, Arizona

CODE/CONDITION*		1986	1987	1988	1989	1990	1991	1992	1993	1994	1995	1996	1997
F02 Cleft lip with and without cleft palate	Cases Rate CI	77 1.25 0.91-1.67	80 1.25 0.92-1.66	91 1.37 1.03-1.79	90 1.33 1.00-1.74	97 1.40 1.06-1.81	80 1.17 0.86-1.55	74 1.07 0.78-1.43	91 1.31 0.98-1.70	83 1.16 0.86-1.53	94 1.29 0.97-1.67	87 1.15 0.86-1.51	103 1.35 1.03-1.73
F08 Pyloric stenosis	Cases Rate CI	108 1.76 1.35-2.25	135 2.11 1.67-2.63	134 2.03 1.60-2.52	122 1.81 1.41-2.27	116 1.68 1.30-2.12	148 2.16 1.73-2.66	137 1.98 1.57-2.46	127 1.82 1.43-2.29	159 2.23 1.80-2.72	148 2.03 1.63-2.50	140 1.85 1.47-2.30	152 1.99 1.60-2.45
F09 TE fistula, esophageal atresia, or both	Cases Rate CI	19 0.31 0.15-0.54	16 0.25 0.11-0.46	19 0.28 0.14-0.50	18 0.27 0.13-0.48	19 0.27 0.14-0.48	15 0.22 0.10-0.41	14 0.20 0.09-0.39	13 0.19 0.08-0.37	14 0.20 0.09-0.38	18 0.25 0.12-0.44	15 0.20 0.09-0.37	21 0.28 0.14-0.47
F14 Stenosis/atresia of duodenum	Cases Rate CI	5 0.08 0.01-0.23	15 0.07 0.01-0.22	11 0.16 0.06-0.34	10 0.15 0.05-0.32	10 0.14 0.05-0.31	6 0.09 0.02-0.23	13 0.19 0.08-0.37	14 0.20 0.09-0.39	14 0.20 0.09-0.38	9 0.12 0.04-0.28	19 0.25 0.13-0.44	11 0.14 0.06-0.30
F15 Stenosis/atresia of small intestine	Cases Rate CI	18 0.29 0.14-0.52	12 0.18 0.07-0.37	13 0.19 0.08-0.38	16 0.24 0.11-0.44	16 0.23 0.11-0.43	9 0.13 0.05-0.29	13 0.19 0.08-0.37	13 0.19 0.08-0.37	15 0.21 0.10-0.40	11 0.15 0.06-0.31	20 0.26 0.14-0.46	12 0.16 0.06-0.32
F16 Stenosis/atresia of rectum or anus	Cases Rate CI	27 0.44 0.25-0.71	26 0.40 0.23-0.66	27 0.40 0.23-0.66	35 0.52 0.32-0.79	35 0.51 0.31-0.78	38 0.56 0.35-0.83	31 0.45 0.27-0.70	27 0.39 0.22-0.63	25 0.35 0.20-0.57	37 0.51 0.32-0.77	29 0.38 0.22-0.61	16 0.21 0.10-0.39
F17 Hirschsprung's Disease	Cases Rate CI	11 0.17 0.07-0.37	10 0.15 0.05-0.33	10 0.15 0.05-0.32	7 0.03 0.03-0.25	13 0.19 0.08-0.37	13 0.19 0.08-0.37	7 0.10 0.03-0.25	8 0.11 0.04-0.27	10 0.14 0.05-0.30	16 0.22 0.10-0.41	9 0.12 0.04-0.27	8 0.10 0.03-0.24

* See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.
"Cases" are the number of live born and stillborn infants $20 weeks gestation.

Table 3 Continued
Congenital Anomalies by Year, 1986 - 1997
Incidence Rates[a] Per 1,000 Live Births and Fetal Deaths, Arizona

CODE/CONDITION *													
		1986	1987	1988	1989	1990	1991	1992	1993	1994	1995	1996	1997
F18 Malrotation of intestine	Cases Rate CI	10 0.16 0.06-0.35	10 0.15 0.05-0.33	16 0.24 0.11-0.44	14 0.21 0.09-0.40	16 0.23 0.11-0.43	14 0.20 0.09-0.39	10 0.14 0.05-0.31	20 0.29 0.15-0.50	24 0.34 0.19-0.56	19 0.26 0.13-0.46	22 0.29 0.16-0.49	16 0.21 0.10-0.39
F21 Biliary atresia	Cases Rate CI	2 0.03 0.00-0.15	1 0.01 0.00-0.11	3 0.04 0.00-0.16	5 0.07 0.02-0.21	4 0.06 0.01-0.18	6 0.09 0.02-0.23	4 0.06 0.01-0.18	8 0.11 0.04-0.27	6 0.08 0.02-0.22	3 0.04 0.00-0.15	7 0.09 0.03-0.23	2 0.03 0.00-0.12
H01 Renal agenesis	Cases Rate CI	21 0.34 0.18	27 0.42 0.24-0.68	23 0.34 0.18-0.58	43 0.64 0.41-0.93	33 0.48 0.29-0.74	37 0.54 0.34-0.82	33 0.48 0.29-0.74	30 0.43 0.25-0.68	25 0.35 0.20-0.57	39 0.54 0.34-0.80	39 0.52 0.33-0.77	34 0.45 0.27-0.68
H06 Obstruction of kidney or ureter	Cases Rate CI	37 0.60 0.37-0.91	71 1.11 0.80-1.50	64 0.97 0.68-1.32	90 1.33 1.00-1.74	94 1.36 1.02-1.76	103 1.50 1.15-1.93	73 1.05 0.76-1.42	73 1.05 0.76-1.41	86 1.20 0.90-1.58	108 1.48 1.14-1.89	115 1.52 1.18-1.93	110 1.44 1.11-1.84
H09 Bladder or urethra obstruction	Cases Rate CI	8 0.13 0.04-0.30	12 0.18 0.07-0.37	9 0.13 0.04-0.30	7 0.10 0.03-0.25	3 0.04 0.00-0.16	8 0.12 0.04-0.27	7 0.10 0.03-0.25	14 0.20 0.09-0.39	11 0.15 0.06-0.32	7 0.10 0.03-0.24	13 0.17 0.07-0.34	10 0.13 0.05-0.28
J03 Dislocation of hip	Cases Rate CI	87 1.42 1.05-1.86	101 1.58 1.20-2.03	68 1.03 1.20-2.03	91 1.35 1.01-1.76	105 1.52 1.16-1.76	103 1.50 1.15-1.93	66 0.95 0.68-1.30	74 1.06 0.77-1.43	65 0.91 0.65-1.24	83 1.14 0.84-1.50	67 0.89 0.63-1.21	55 0.72 0.50-1.01
J51 Complete absence of upper or lower limb	Cases Rate CI	2 0.03 0.00-0.15	0 0.00 0.00-0.00	1 0.01 0.00-0.11	3 0.04 0.00-0.16	3 0.04 0.00-0.16	2 0.03 0.00-0.14	1 0.01 0.00-0.11	2 0.03 0.00-0.13	1 0.01 0.00-0.10	2 0.03 0.00-0.13	3 0.04 0.00-0.15	0 0.00 0.00-0.00

* See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.
“Cases” are the number of live born and stillborn infants $20 weeks gestation.

Table 3 Continued
Congenital Anomalies by Year, 1986 - 1997
Incidence Rates[a] Per 1,000 Live Births and Fetal Deaths, Arizona

CODE/CONDITION*													
		1986	1987	1988	1989	1990	1991	1992	1993	1994	1995	1996	1997
J52 Phocomelia of limb	Cases Rate CI	3 0.04 0.00-0.18	2 0.03 0.00-0.14	2 0.03 0.00-0.14	1 0.01 0.00-0.25	1 0.01 0.00-0.11	1 0.01 0.00-0.11	0 0.00 0.00-0.00	1 0.01 0.00-0.11	2 0.03 0.00-0.13	0 0.00 0.00-0.00	0 0.00 0.00-0.00	0 0.00 0.00-0.00
K05 Amniotic bands	Cases Rate CI	4 0.06 0.01-0.20	4 0.06 0.01-0.19	9 0.14 0.05-0.32	8 0.11 0.04-0.28	14 0.20 0.09-0.39	10 0.15 0.05-0.31	8 0.12 0.04-0.27	7 0.10 0.03-0.25	9 0.13 0.04-0.28	12 0.16 0.07-0.33	6 0.08 0.02-0.21	12 0.16 0.06-0.32
N01 Diaphragmatic hernia	Cases Rate CI	13 0.21 0.09-0.41	18 0.28 0.13-0.50	20 0.30 0.15-0.52	23 0.34 0.19-0.57	28 0.40 0.23-0.65	23 0.34 0.18-0.56	13 0.19 0.08-0.37	18 0.26 0.13-0.46	21 0.29 0.15-0.50	20 0.27 0.14-0.48	15 0.20 0.09-0.37	15 0.20 0.09-0.37
N02 Omphalocele	Cases Rate CI	10 0.16 0.06-0.35	14 0.21 0.09-0.42	17 0.25 0.12-0.46	10 0.15 0.05-0.32	21 0.30 0.16-0.52	21 0.31 0.16-0.53	10 0.14 0.05-0.31	17 0.24 0.12-0.44	11 0.15 0.06-0.32	14 0.19 0.09-0.37	20 0.26 0.14-0.46	11 0.14 0.06-0.30
N04 Gastroschisis	Cases Rate CI	19 0.31 0.15-0.54	18 0.28 0.13-0.50	19 0.28 0.14-0.50	19 0.28 0.14-0.50	21 0.30 0.16-0.52	36 0.53 0.33-0.80	27 0.39 0.22-0.63	16 0.23 0.11-0.42	27 0.38 0.22-0.61	27 0.37 0.21-0.60	42 0.56 0.36-0.82	36 0.47 0.29-0.72
R01 Down Syndrome (Trisomy 21)	Cases Rate CI	64 1.04 0.73-1.43	61 0.95 0.67-1.32	74 1.12 0.81-1.50	66 0.98 0.70-1.33	73 1.05 0.76-1.42	84 1.23 0.91-1.62	87 1.26 0.94-1.65	80 1.15 0.84-1.52	100 1.40 1.07-1.80	90 1.23 0.92-1.61	95 1.26 0.95-1.63	101 1.33 1.01-1.71
R02 Patau Syndrome (Trisomy 13)	Cases Rate CI	9 0.14 0.05-0.32	4 0.06 0.01-0.19	3 0.04 0.00-0.16	4 0.06 0.01-0.19	11 0.16 0.06-0.33	6 0.09 0.02-0.23	15 0.22 0.10-0.41	9 0.13 0.04-0.29	3 0.04 0.00-0.15	8 0.11 0.03-0.26	13 0.17 0.07-0.34	5 0.07 0.01-0.19

* See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.
“Cases” are the number of live born and stillborn infants $20 weeks gestation.

Table 3 Continued
Congenital Anomalies by Year, 1986 - 1997
Incidence Rates Per 1,000 Live Births and Fetal Deaths[1], Arizona

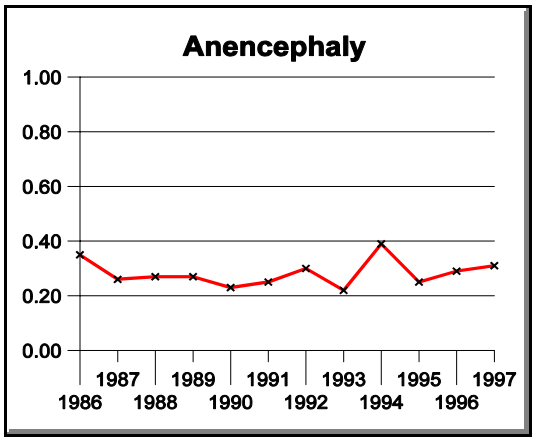
CODE/CONDITION*													
		1986	1987	1988	1989	1990	1991	1992	1993	1994	1995	1996	1997
R03 Edwards Syndrome (Trisomy 18)	Cases Rate CI	11 0.17 0.07-0.37	17 0.26 0.12-0.48	13 0.19 0.08-0.38	10 0.15 0.05-0.32	15 0.22 0.10-0.41	13 0.19 0.08-0.37	12 0.17 0.07-0.35	14 0.20 0.09-0.39	14 0.20 0.09-0.38	18 0.25 0.12-0.44	16 0.21 0.10-0.39	15 0.20 0.09-0.37
S02 Fetal alcohol syndrome	Cases Rate CI	9 0.14 0.05-0.32	25 0.39 0.21-0.64	12 0.18 0.07-0.36	21 0.31 0.16-0.53	22 0.32 0.17-0.54	27 0.39 0.23-0.64	33 0.48 0.29-0.74	26 0.37 0.21-0.61	14 0.20 0.09-0.38	27 0.37 0.21-0.60	10 0.13 0.05-0.28	9 0.12 0.04-0.26

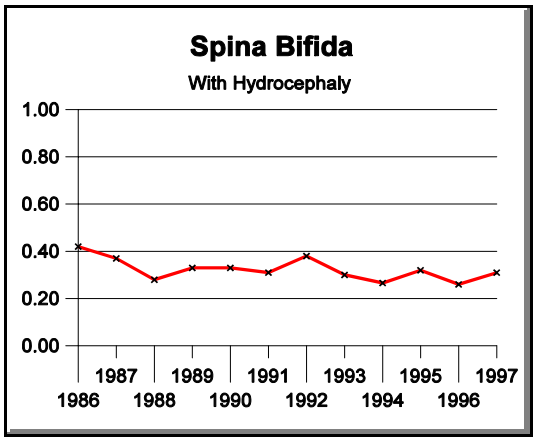
* See appendix for explanation of the codes/conditions
CI = Approximate 99% confidence intervals.
“Cases” are the number of live born and stillborn infants $20 weeks.

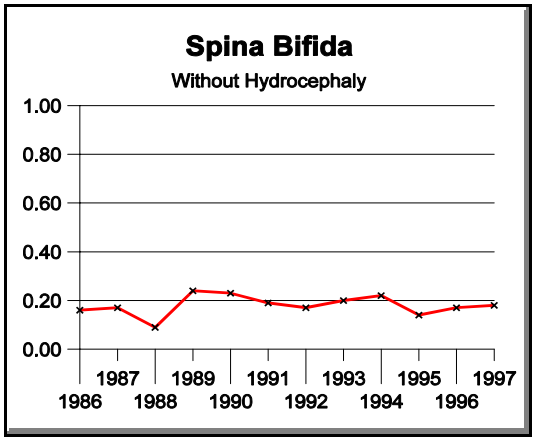
[a] The rates are calculated as the number of live born and stillborn cases of each defect divided by the denominators consisting of the total live births and stillbirths as follows:

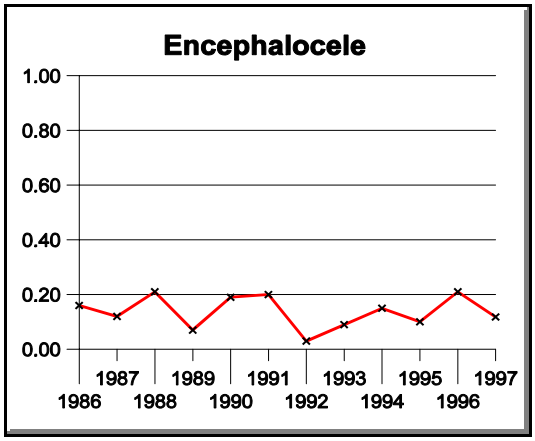
1986 = 61,203; 1987 = 63,742; 1988 = 65,981; 1989 = 67,498; 1990 = 69,245; 1991 = 68,449; 1992 = 69,202; 1993=69,593; 1994 = 71,403; 1995 = 72,883; 1996=75,577; 1997 = 76,200.

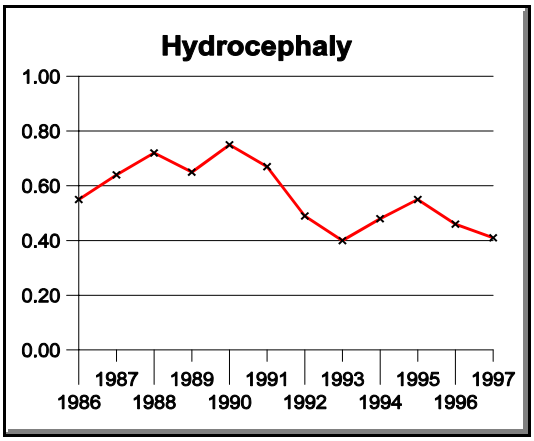
Figure 2
Trends of Selected Congenital Anomalies, Incidence Rates, 1986-1997
(Live Born and Stillborn Cases Per 1,000 Live Births & Fetal Deaths), Arizona











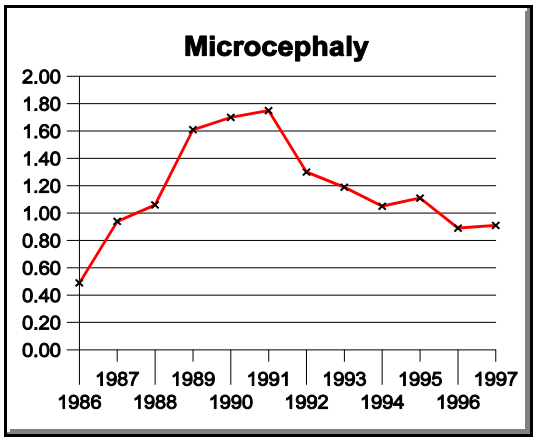
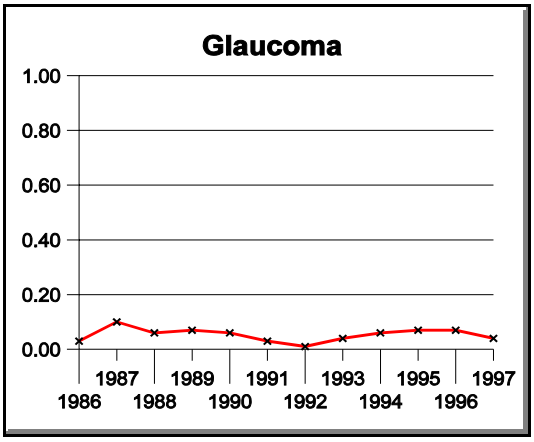
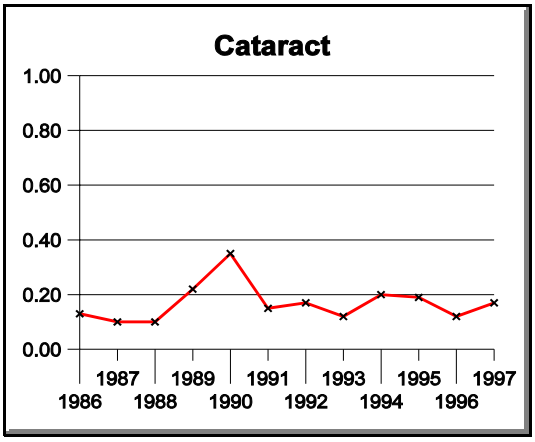
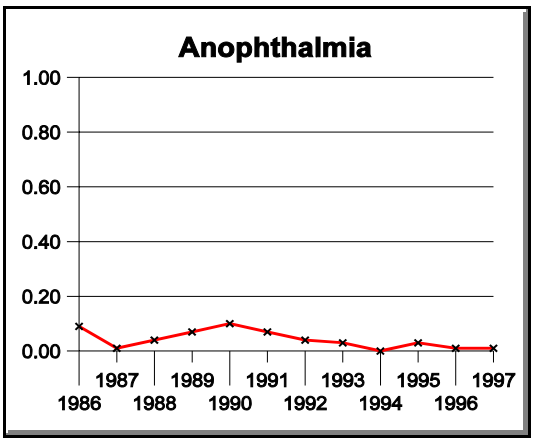
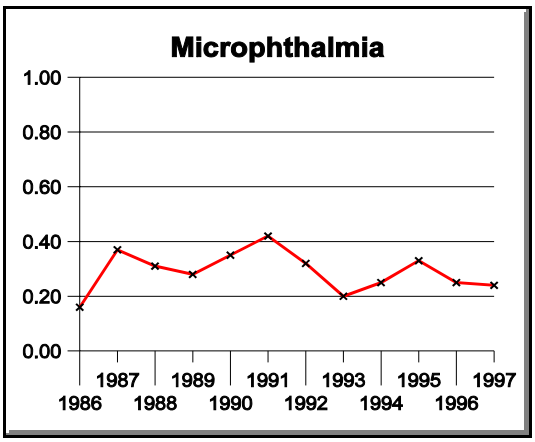


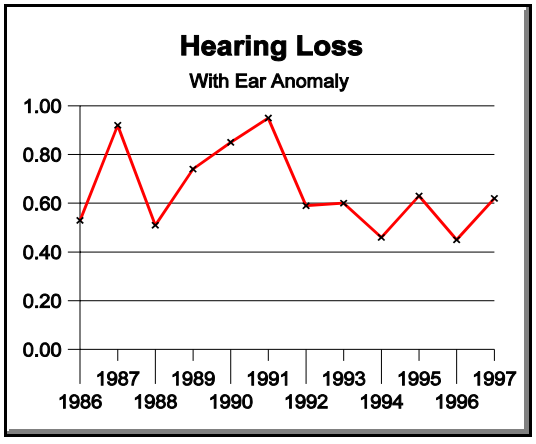
Figure 2 Continued
Trends of Selected Congenital Anomalies: Incidence Rates
(Live Born and Stillborn Cases Per 1,000 Live Births & Fetal Deaths), Arizona











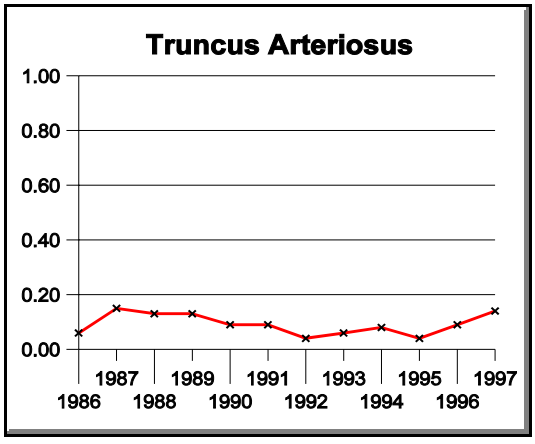
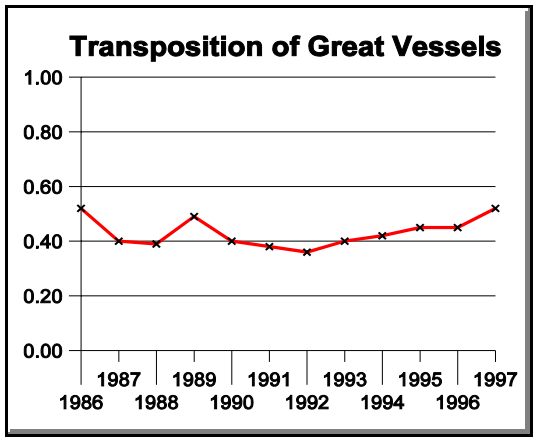
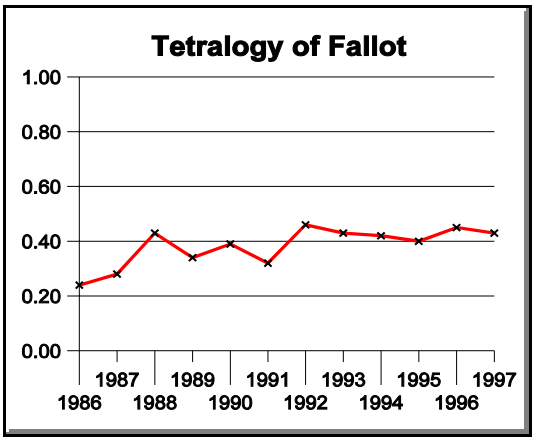
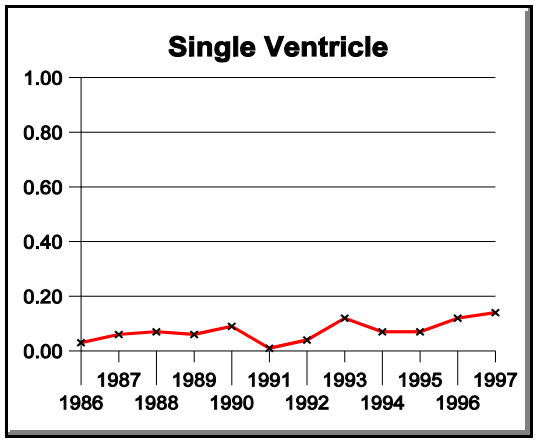
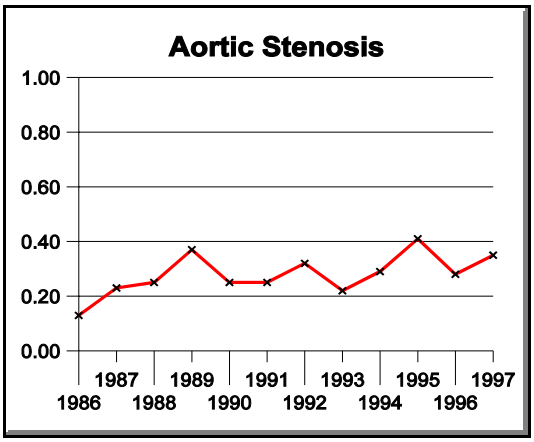


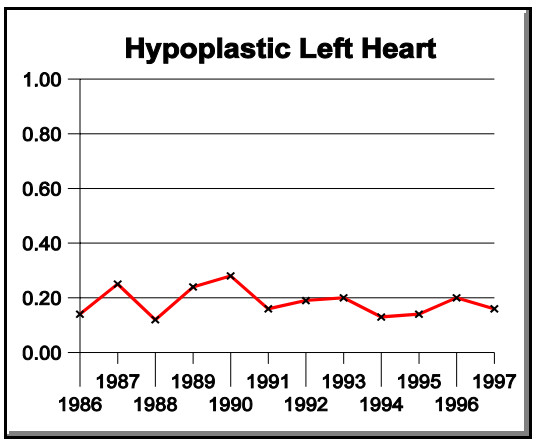
Figure 2 Continued
Trends of Selected Congenital Anomalies: Incidence Rates
(Live Born and Stillborn Cases Per 1,000 Live Births & Fetal Deaths), Arizona











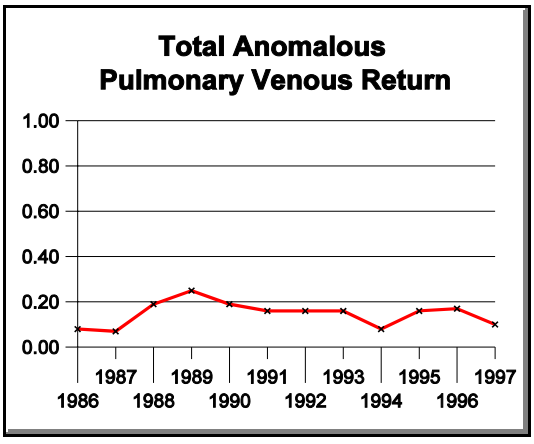
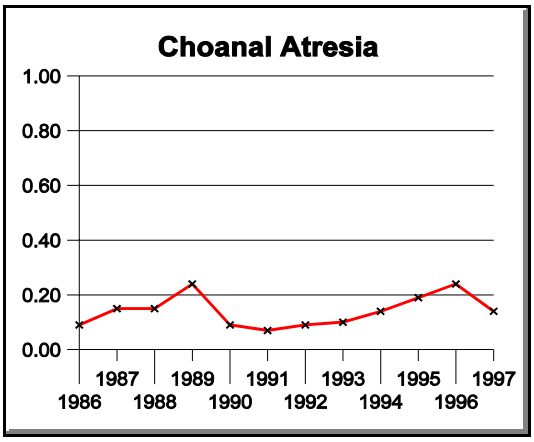
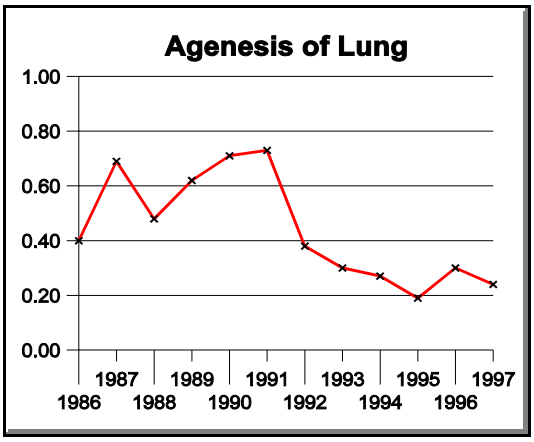
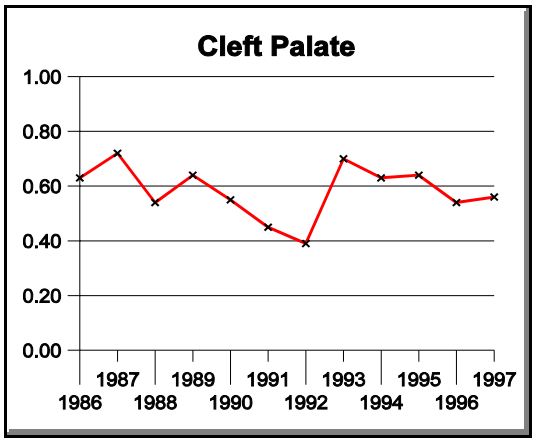
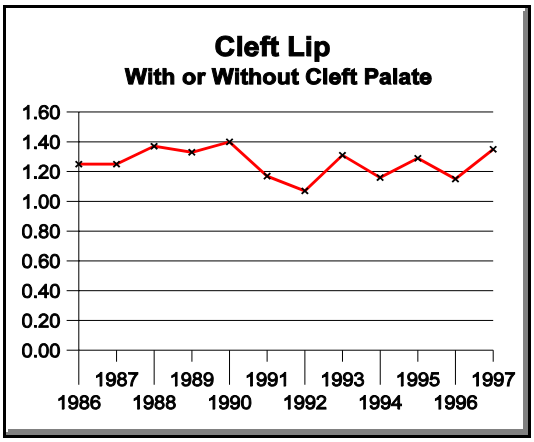


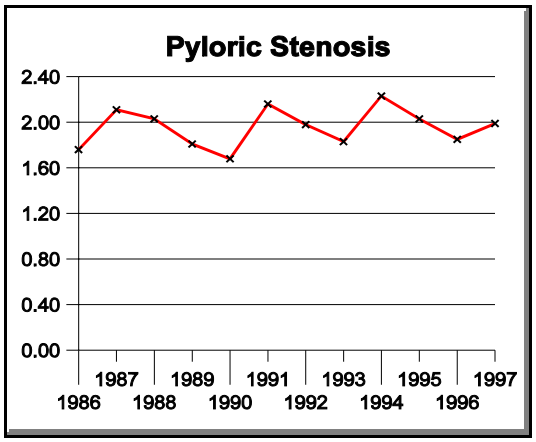
Figure 2 Continued
Trends of Selected Congenital Anomalies: Incidence Rates
(Live Born and Stillborn Cases Per 1,000 Live Births & Fetal Deaths), Arizona











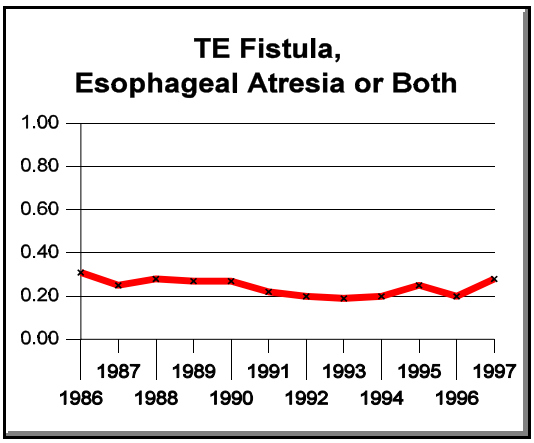
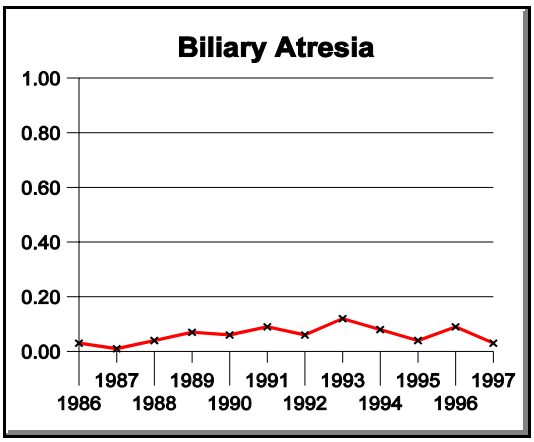
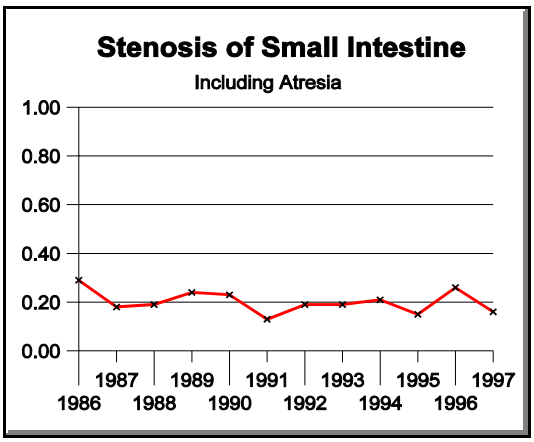
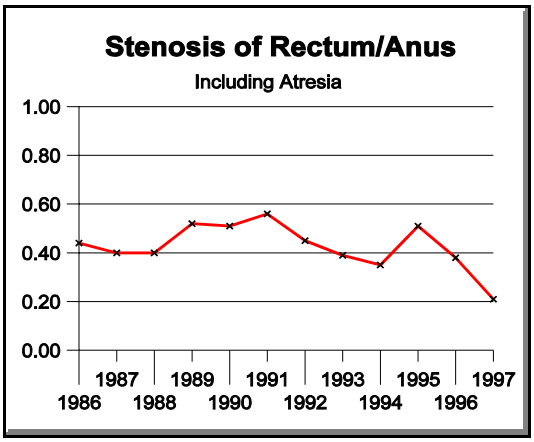
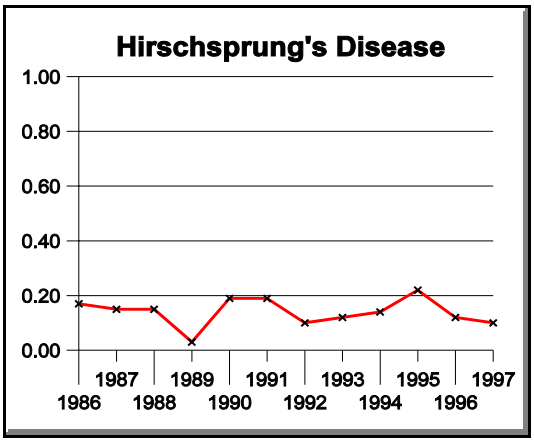


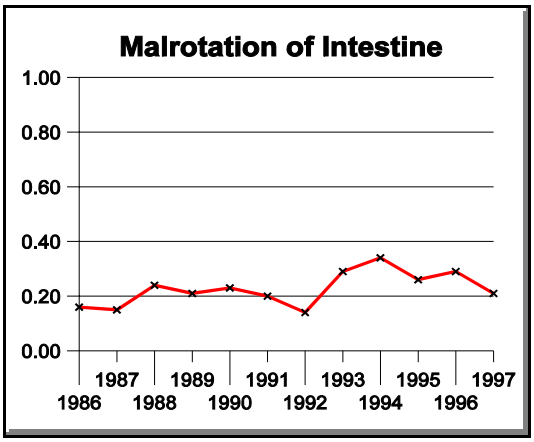
Figure 2 Continued
Trends of Selected Congenital Anomalies: Incidence Rates
(Live Born and Stillborn Cases Per 1,000 Live Births & Fetal Deaths), Arizona











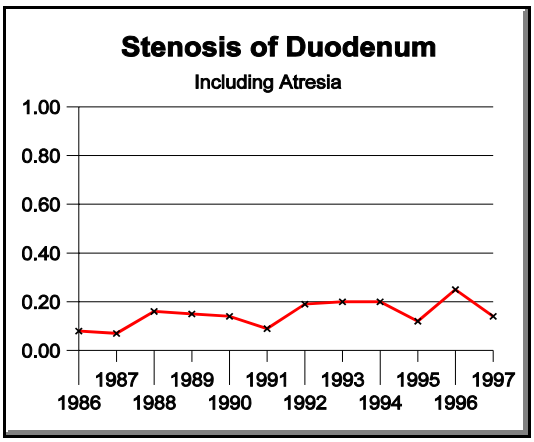
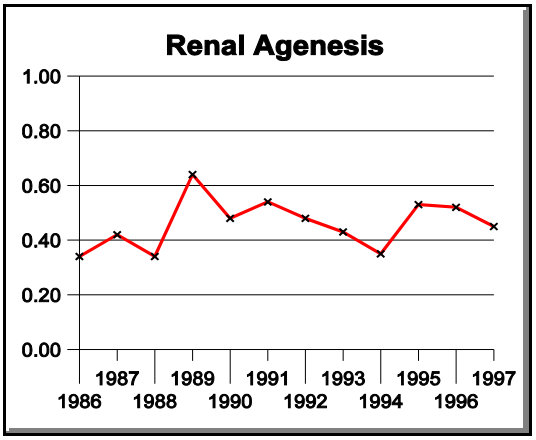
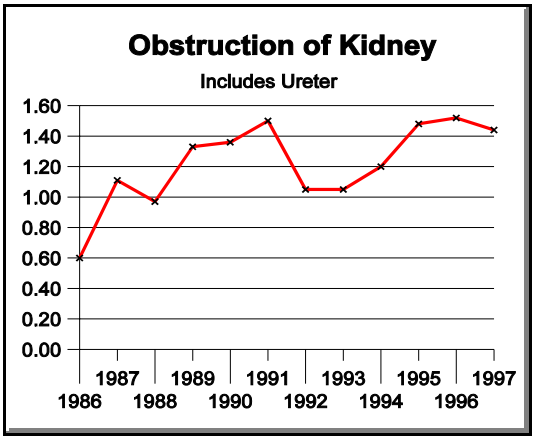
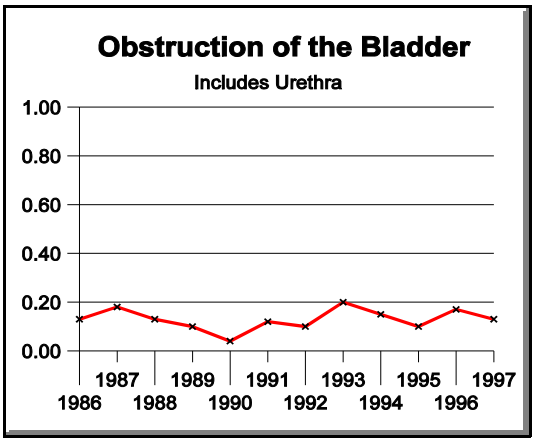
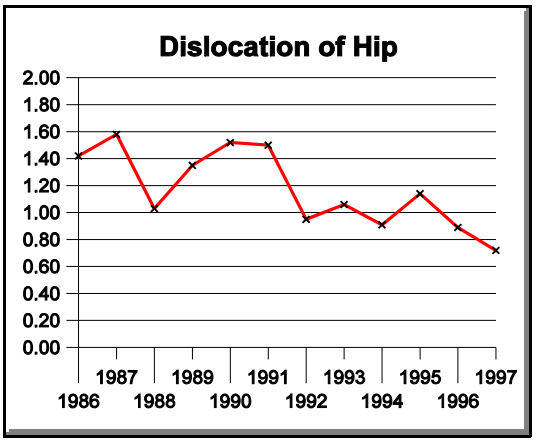


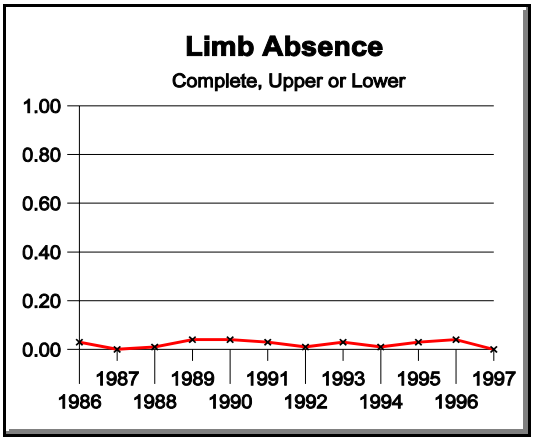
Figure 2 Continued
Trends of Selected Congenital Anomalies: Incidence Rates
(Live Born and Stillborn Cases Per 1,000 Live Births & Fetal Deaths), Arizona











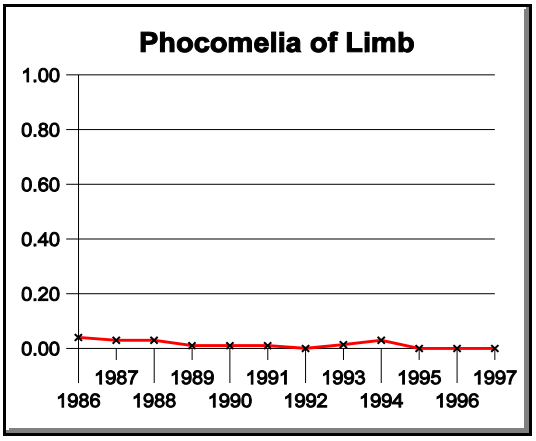
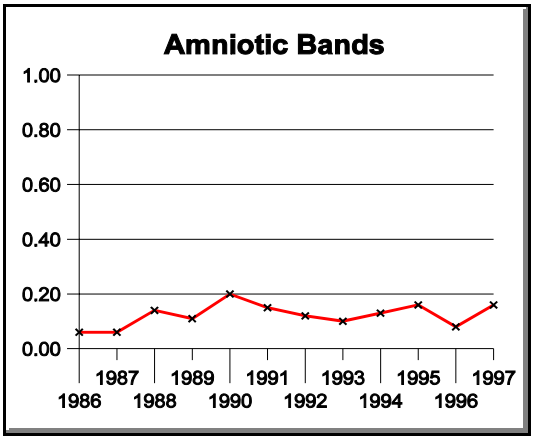
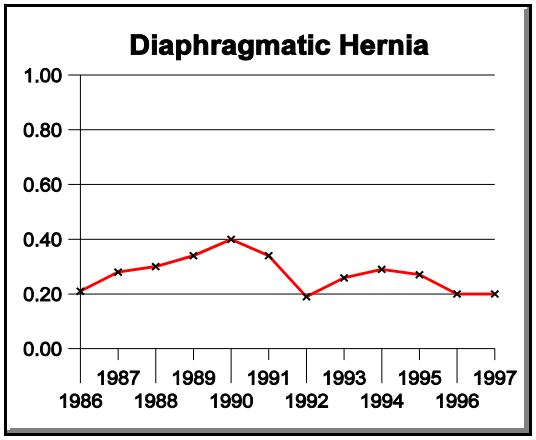
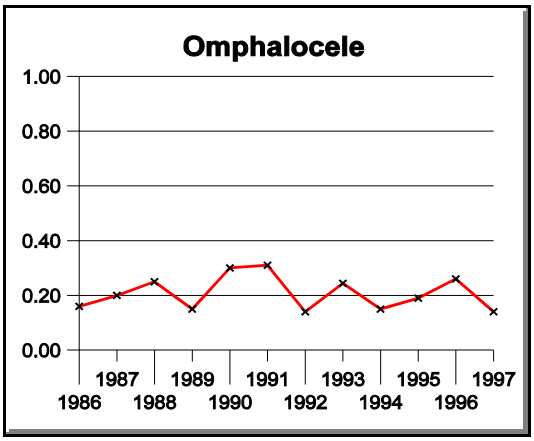
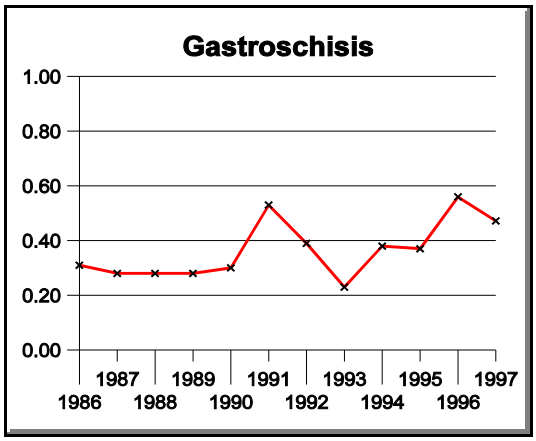


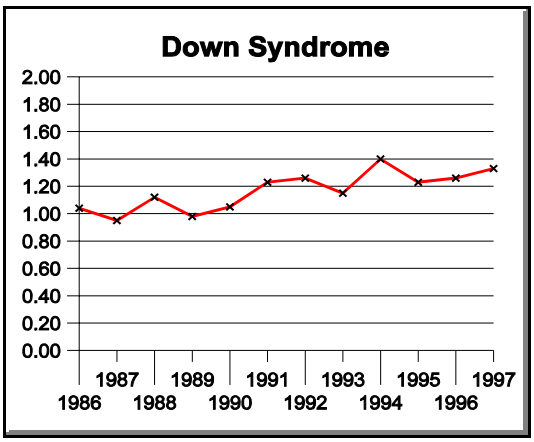
Figure 2 Continued
Trends of Selected Congenital Anomalies: Incidence Rates
(Live Born and Stillborn Cases Per 1,000 Live Births & Fetal Deaths), Arizona











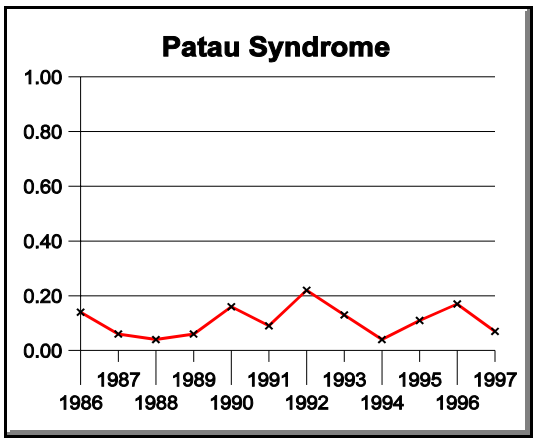
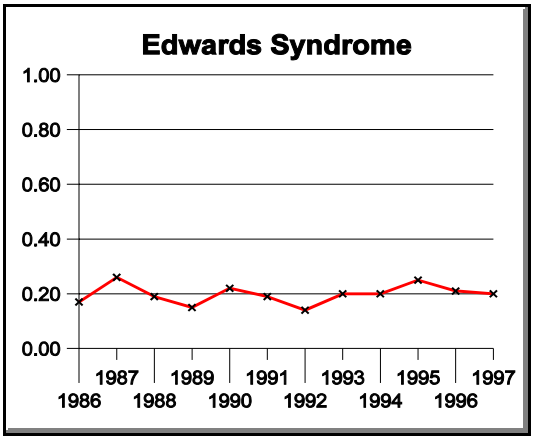
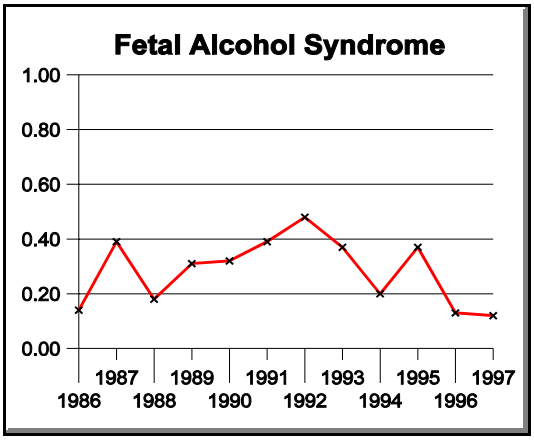


Figure 2 Continued
Trends of Selected Congenital Anomalies: Incidence Rates
(Live Born and Stillborn Cases Per 1,000 Live Births & Fetal Deaths), Arizona





RACE/ETHNICITY

All race and ethnic groups experience birth defects, but the frequency and types of these vary by race and ethnicity.[12,13] The race and ethnicity information collected in the Arizona birth, death and fetal death certificates allow for the analysis of birth defects by race and ethnicity (Appendix 6). The following figures display the rates of selected birth defects by race and ethnicity. However, due to the small number of cases of specific birth defects among the subgroups, the rates for all of the race/ethnic groups are not displayed. Tables 1-A and 1-B show the counts used for the calculation of the rates.

The overall rate for spina bifida (including cases with and without hydrocephaly) was highest among Hispanics in 1997, but was not statistically significant (Figure 3). This is consistent with the literature that indicates that higher rates of spina bifida occur among Hispanics than Whites. A California study suggested that, while the risk for an NTD-affected pregnancy was twice as high among women of Mexican descent compared to Whites, the risk was not substantially higher between U.S. born women of Mexican and other racial/ethnic descent and Whites.[14] A Colorado study further suggests that low maternal education, particularly among Hispanics, was a strong predictor of having a child with an NTD.[15] Due to small numbers of cases occurring among other races, rate comparisons of NTDs were limited to Whites and Hispanics.

Data in Table 1-B (1997) show the same patterns observed in 1996. Rates of abdominal wall defects (gastroschesis and omphalocele) among Hispanics were higher than the rates for Whites (8.52 v. 3.93 per 10,000 live and still births). An examination of the specific defects show that omphalocele rates in 1997 were higher for Whites, relative to Hispanics. However, gastroschisis rates were higher among Hispanics, relative to Whites (Figure 4) that year. Tests of significance show that gastroschisis rates for Hispanics were not significantly different from the state rate of 4.72 per 10,000 live births and fetal deaths in 1997.

Down Syndrome (Trisomy 21) rates in 1997 were highest for Blacks, at 23.87, followed by Native Americans, at 22.56, Hispanics, at 13.49, and Whites, at 11.52 per 10,000 live and stillbirths. Although rates for Blacks, Native Americans and Hispanics were higher than the state rate of 13.25 per 10,000 live and stillbirths, the differences in these rates were not statistically significant.

Native Americans and Blacks exhibited the highest rates for microcephaly in 1997 (Figure 6). Whites are found to have the highest rates of pyloric stenosis at 23.04 per 10,000 live born and stillborn infants, compared to the overall state rate (19.95 per 10,000 live born and stillborn infants). The next highest rates are among Native Americans (22.56 per 10,000 live born and stillborn infants) (Figure 7). Figure 8 shows that Native Americans have significantly higher rates than the overall state rate for cleft lip with and without cleft palate (41.35 vs. 13.52 per 10,000 live born and stillborn infants at $p < .01$). Fetal alcohol syndrome rates are highest among Native Americans at 11.28 per 10,000 live births and stillborn infants (Figure 9). Z-test results show a significant difference between the Native American fetal alcohol syndrome rate and the state rate at $p < .01$.

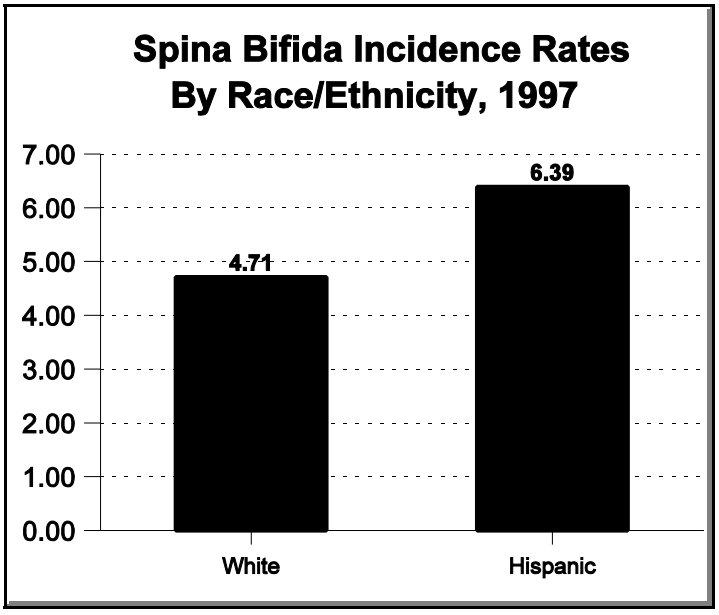


Figure 3. Spina Bifida with and without Hydrocephaly, Incidence Rates (Live Born and Stillborn Cases Per 10,000 Live Births and Fetal Deaths) by Race/Ethnicity, 1997

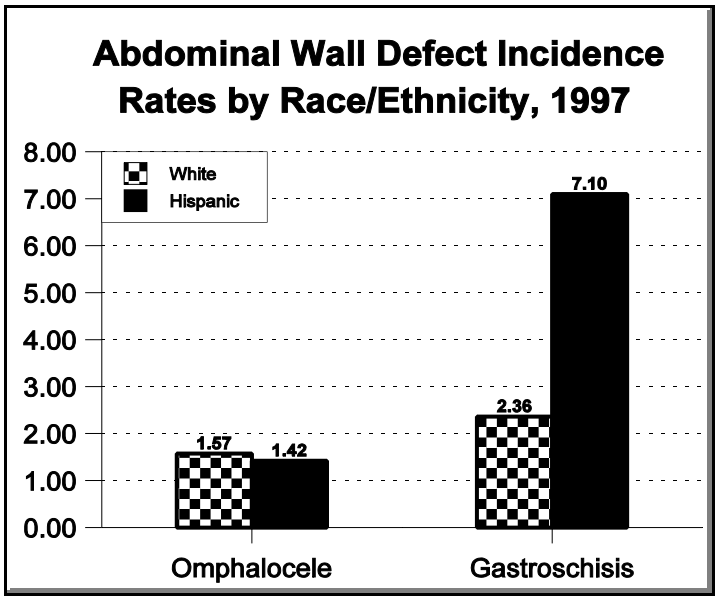


Figure 4. Abdominal Wall Defect Incidence Rates
(Live Born and Stillborn Cases Per 10,000 Live Births and Fetal Deaths)
by Race/Ethnicity, 1997

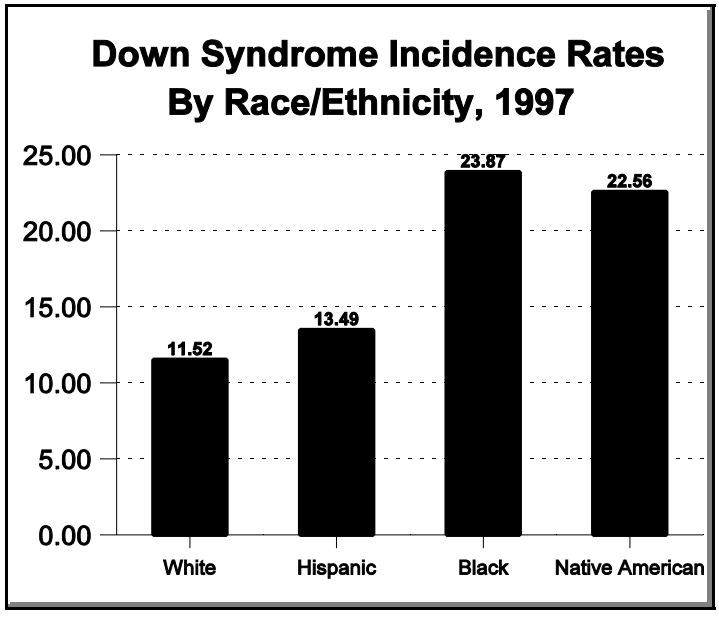


Figure 5. Down Syndrome Incidence Rates
(Live Born and Stillborn Cases Per 10,000 Live Births and Fetal Deaths)
by Race/Ethnicity, 1997

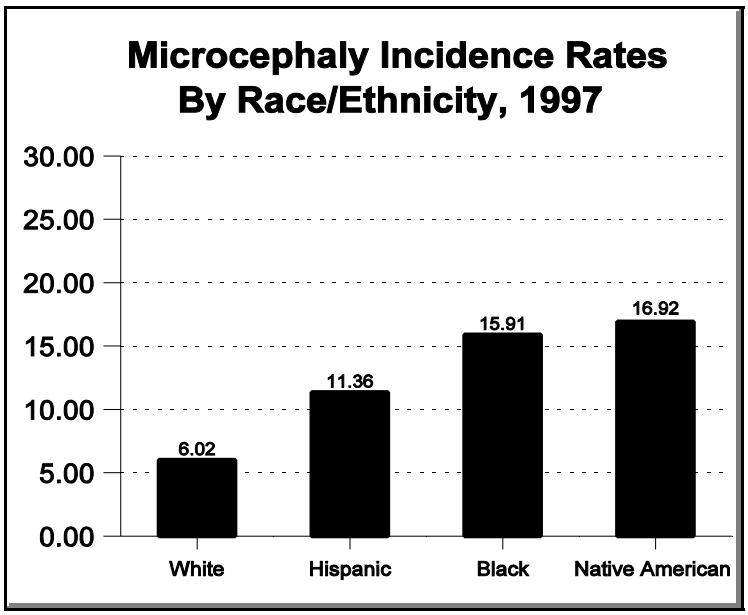


Figure 6. Microcephaly Incidence Rates
(Live Born and Stillborn Cases Per 10,000 Live Births and Fetal Deaths)
by Race/Ethnicity, 1997

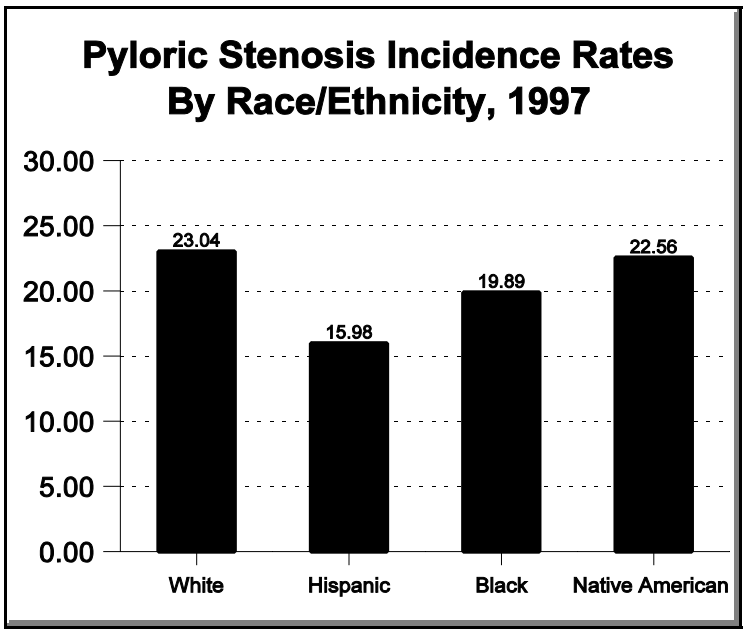


Figure 7. Pyloric Stenosis Incidence Rates (Live Born and Stillborn Cases Per 10,000 Live Births and Fetal Deaths) by Race/Ethnicity, 1997

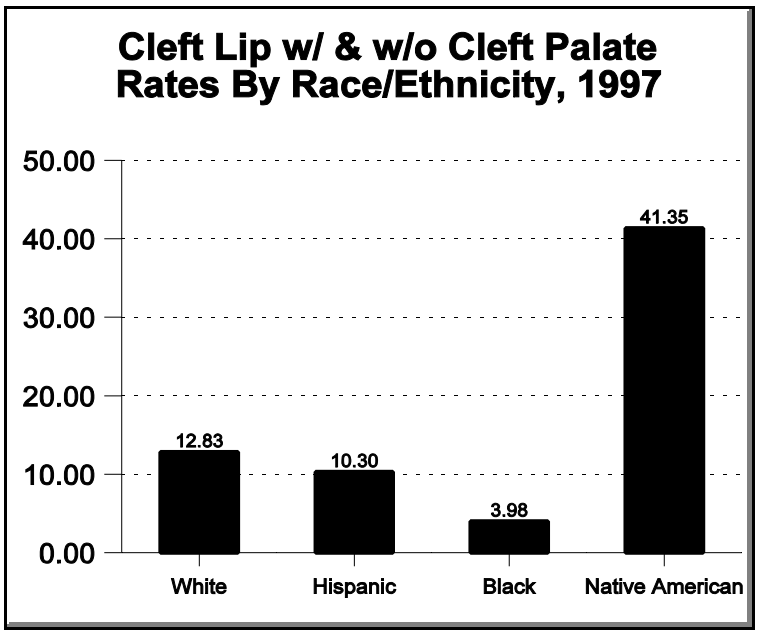


Figure 8. Cleft Lip with and without Cleft Palate Incidence Rates (Live Born and Stillborn Cases Per 10,000 Live Births and Fetal Deaths) by Race/Ethnicity, 1997

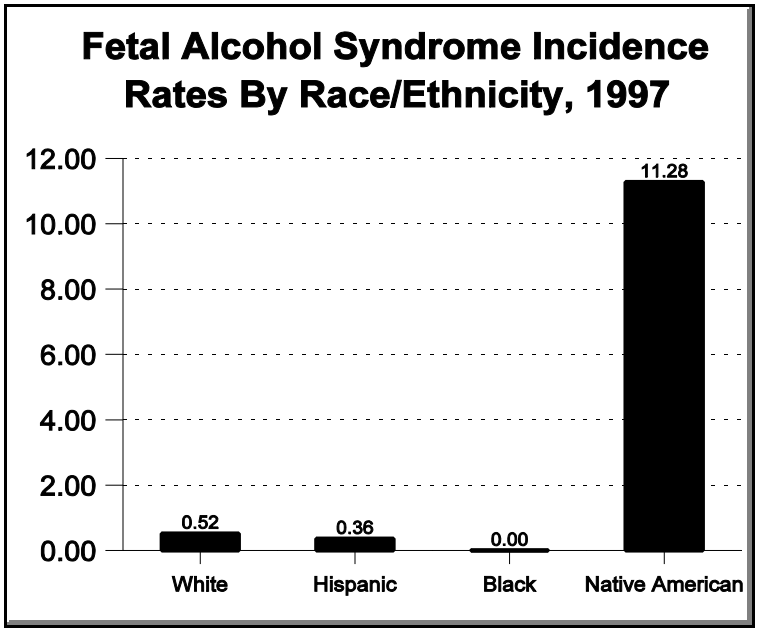


Figure 9. Fetal Alcohol Syndrome Incidence Rates (Live Born and Stillborn Cases Per 10,000 Live Births and Fetal Deaths) by Race/Ethnicity, 1997

MATERNAL AGE

Maternal age was divided into five age groups. Observed rates of the 44 categories of congenital anomalies collected were highest among women 35 years of age and older, followed by the <20-year-old age group, in both 1994 and 1997. In 1997 the birth defect rates for mothers who were in both of these age categories were significantly higher than the state rate ($p < .01$). In both years the rates for birth defects were lowest among the 25- to 29-year-old maternal age group. Z-test results show that birth defect rates for women who were 35 years of age and older were significantly higher than the state rate at $p < .01$ for these years. In particular, Down Syndrome (Trisomy 21) rates increased with maternal age (Figure 11). The incidence of Down Syndrome for mothers 35 years and older was significantly higher than the state rate for Down Syndrome, and the incidence rate for the 25- to 29-year-old maternal age group was significantly lower than the state rate for the syndrome in 1994 and 1997. In contrast, rates for gastroschisis decreased with increasing maternal age (Figure 12). However, none of the rates of gastroschisis for specific maternal age groups were statistically different from the state rate for gastroschisis.

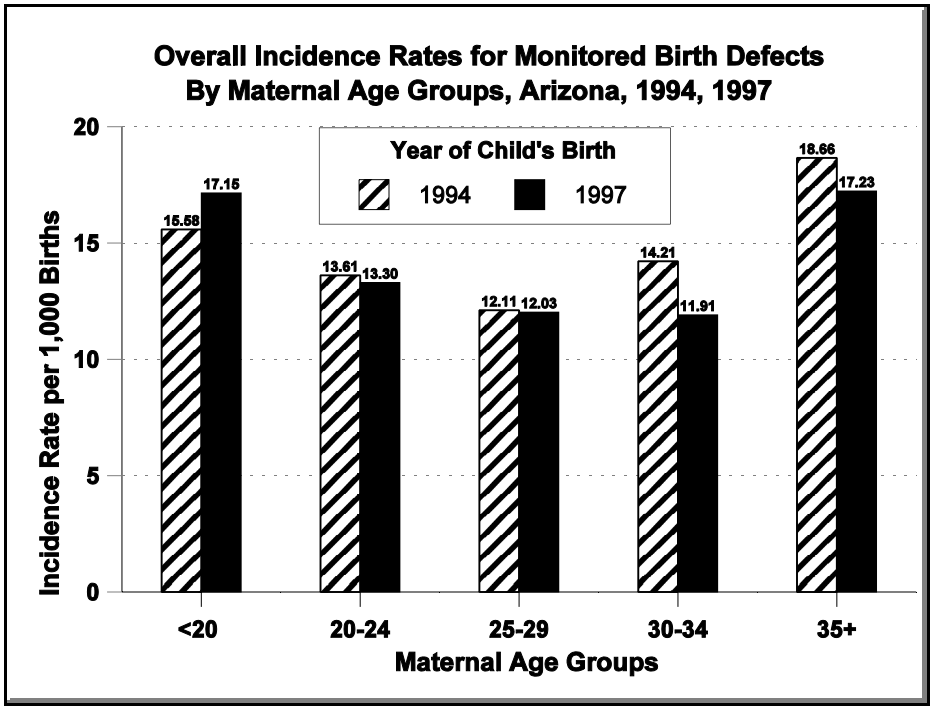


Figure 10. Overall Incidence Rates for the 44 Monitored Defects Listed on Tables 1-A and B. By Maternal Age Groups, Arizona, 1994 and 1997
(Live Born and Stillborn Cases Per 1,000 Live Births and Fetal Deaths)

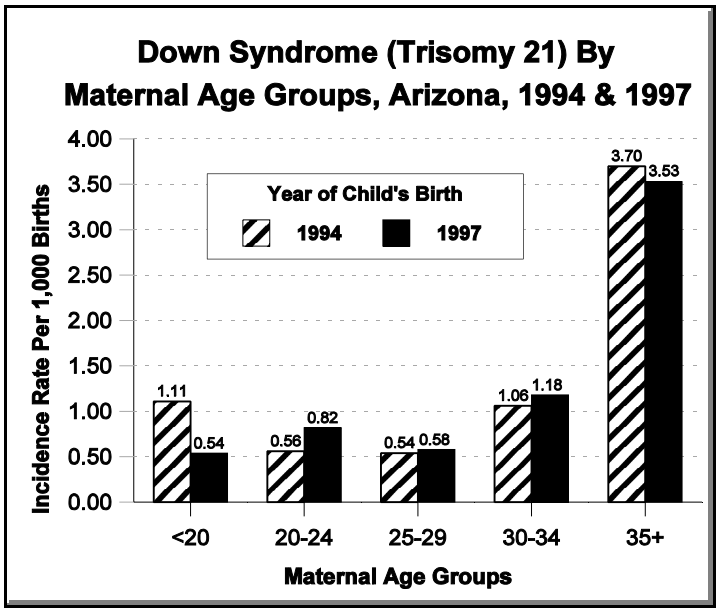


Figure 11. Down Syndrome (Trisomy 21) Incidence Rates, 1994 & 1997
(Live Born and Stillborn Cases Per 1,000 Live Births and Fetal Deaths)
by Maternal Age Groups

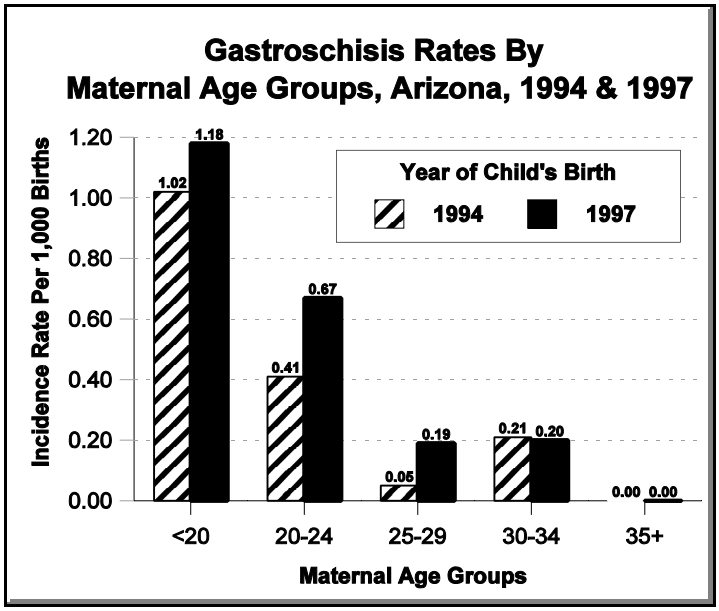


Figure 12. Gastroschisis Incidence Rates, 1994 & 1997
(Live Born and Stillborn Cases Per 1,000 Live Births and Fetal Deaths)
by Maternal Age Groups

COUNTY PROFILES

Using County Data

The Arizona Birth Defect Monitoring Program (ABDMP) collects birth defect information from all of Arizona's 15 counties. Multiple years are used to give sufficient data to derive statistically stable measures at the county level. Birth defect data from live births are analyzed in this section.

Dealing With Small Numbers

Analysis of county data is difficult because of normal fluctuations in rates seen in small populations. When dealing with small numbers, it is normal to see fluctuations over time. With rate fluctuations we may see the appearance of birth defect clusters. Most often this is a statistical anomaly. In the rare case that a cluster results from a newly introduced teratogen, a dramatic increase on the scale of 10-fold or greater is usually seen.[17] Small numbers are also withheld to help protect the children and their parents' confidentiality. Thus, all county level data are aggregated. Incidence rates and confidence intervals are presented when there are 10 or more cases. Z-tests were used to test for the equivalence between the county rates and the state rate.

Birth Defects by County

The following tables present birth defects by county of mothers' residences. Cases were aggregated for the years 1986 through 1997. Table 4 shows the total cases and rates of the 44 categories of congenital anomalies collected for each Arizona county. Gila and Navajo Counties had the highest rates of infants with congenital anomalies, at 17.12 and 15.79 per 1,000 live births. Statistical analysis indicates that these county rates are significantly different form the state rate of 13.16 per 1,000 live births. Table 5 examines the 44 categories of anomalies by race and county. For Whites, Maricopa County has the highest rate of birth defects, followed by Pima County. For Hispanics, Navajo County has the highest rate of congenital anomalies, and for Blacks, Pima County has the highest rate. Yuma and Graham Counties have the highest rates for Native Americans. The rate for each race/ethnicity group for each county is compared with that of the state rate to assess whether the county rates for each of the race/ethnic groups are significantly different from the state rate. The Z-test results indicate that Yuma and Graham Counties' Native American rates are significantly higher than the state rate, with 99 percent confidence. Other county rates for the other race/ethnic groupings were not significantly different from the state rate.

Table 4
Birth Defect Incidence Rates* by County 1986-1997
(Live Born Cases Per 1,000 Live Births)

COUNTY	CASES 1986-1997	RATE	99% CONFIDENCE INTERVAL
Arizona	10,861	13.16	12.88-13.64
Apache	267	13.72	11.65-16.04
Cochise	218	10.85	9.05-12.89
Coconino	298	13.17	11.28-15.27
Gila	136	17.12	13.57-21.28
Graham	75	14.33	10.42-19.18
Greenlee	12	7.33	3.00-14.78
La Paz	23	9.81	5.33-16.43
Maricopa	6,432	13.13	12.71-13.55
Mohave	204	11.08	9.18-13.24
Navajo	350	15.79	13.70-18.10
Pima	1,781	13.23	12.44-14.06
Pinal	358	14.28	12.41-16.35
Santa Cruz	114	12.92	10.01-16.38
Yavapai	185	11.61	9.53-14.00
Yuma	408	13.18	11.56-14.96

*Overall incidence rates for the 44 categories of birth defects monitored (see Tables 1-A and B)

Table 5
Birth Defect Incidence Rates by Race/Ethnicity, by County, 1986-1997
(Live Born Cases Per 1,000 Live Births)

COUNTY	WHITE NON-HISP.	HISPANIC	BLACK	NATIVE AMERICAN	OTHER
	Rate 99% C.I.	Rate 99% C.I.	Rate 99% C.I.	Rate 99% C.I.	Rate 99% C.I.
Arizona	11.93 11.52-12.36	13.58 13.00-14.18	12.19 10.71-13.81	17.48 16.20-18.82	9.58 7.61-11.89
Apache	9.09 4.78-15.59	15.90 5.86-34.09	- -	14.26 11.98-16.84	- -
Cochise	10.41 7.99-13.30	10.88 8.09-14.29	9.72 4.58-17.94	- -	- -
Coconino	10.04 7.60-13.01	12.15 7.26-19.01	- -	15.46 12.47-18.94	- -
Gila	11.70 7.69-17.01	11.67 6.14-20.01	- -	26.00 18.21-35.89	- -
Graham	11.68 7.16-17.92	12.70 6.68-21.77	- -	26.72 13.52-47.01	- -
Greenlee	- -	- -	- -	- -	- -
La Paz	9.53 3.51-20.44	- -	- -	- -	- -
Maricopa	12.26 11.74-12.78	14.03 13.25-14.85	11.83 10.10-13.75	18.90 16.03-22.11	9.23 6.63-11.68
Mohave	10.80 8.75-13.19	10.84 6.21-17.49	- -	- -	- -
Navajo	10.87 7.81-14.70	18.60 10.65-29.99	- -	17.66 14.91-20.76	- -
Pima	11.91 10.87-13.02	13.24 12.00-14.56	15.02 11.30-19.97	20.14 15.60-25.54	14.28 5.55-15.64
Pinal	11.81 9.35-14.69	13.21 10.39-16.53	- -	22.69 16.64-30.15	- -
Santa Cruz	9.02 3.69-18.19	12.93 9.86-16.61	- -	- -	- -
Yavapai	11.62 9.33-14.29	12.03 7.11-18.96	- -	- -	- -
Yuma	10.85 8.43-13.73	13.62 11.57-15.93	- -	30.30 11.82-62.88	- -

- = Insufficient cases for rate and confidence interval calculations.

SENTINEL DEFECTS

Tables 6-10 look at the following sentinel defects: chromosomal defects, oro-facial clefts, neural tube defects, abdominal wall defects, and heart defects. These defects were chosen because of their significant public health impact.

Chromosomal Defects

Chromosomal abnormalities may arise from a deletion (monosomy) or an addition (trisomy) of genetic components that result in various levels of abnormal physical features, structural defects, mental retardation, fetal and infant death, and shortened life expectancy. The most common chromosomal defect is Down Syndrome. Table 6 presents information on Down Syndrome, as well as two other chromosomal trisomies: Patau Syndrome and Edwards Syndrome. Research shows that the risk of a trisomy-affected pregnancy increases with maternal age; however, ethnic differences may impact this risk.[18] There are also data which indicate that about 20 percent of instances of Down Syndrome are paternal in origin. Table 6 shows that rates for chromosomal defects were highest for Santa Cruz County (2.04 per 1,000 live births), followed by Gila County at 1.89 per 1,000 live births. Comparisons of the county rates with the state rate (1.36 per 1,000 live births) for chromosomal defects indicate that there were no significant differences between the county rates and that of the state.

Oral Clefts

Cleft palate is a failure of the palate to fuse properly, forming a grooved fissure in the roof of the mouth. Cleft lip is a failure of the maxillary and median nasal processes to fuse, leaving a fissure in the lip. This occurs between the fourth and eighth week of pregnancy. Babies born with oral clefts may experience problems with eating, drinking, hearing and speech development, requiring services such as surgery and speech therapy. There is evidence that indicates that maternal smoking and alcohol consumption during early pregnancy are associated with increased risks for women having children with oral clefts.[19,20] Table 7 shows that Graham, Apache and Navajo Counties had the highest rates for oral clefts, at 3.06, 3.03 and 2.80 per 1,000 live births, respectively. Greenlee (data not shown), La Paz (data not shown), Mohave and Maricopa Counties had the lowest rates. The state rate for oral clefts was 1.78 per 1,000 live births from 1986 to 1997. Z-tests comparing the rates of oral clefts of the counties with the state rate show that there were no statistical differences between these rates.

Neural Tube Defects

Neural tube defects (NTDs) result from the failure of the neural tube to close properly in fetal development. The two major NTDs are anencephaly and spina bifida. Anencephaly is an absence of part or all of the brain. Spina bifida is a defective closure of the bones of the spine, through which the spinal cord and meninges may or may not protrude. Research indicates that maternal obesity, socioeconomic status and neighborhood social conditions, prior spontaneous and elective terminations, and short periods of time between pregnancies are associated with an increased risk for an NTD-affected pregnancy.[21,22,23] There is, however, ample epidemiologic evidence which shows that daily intake of 400 mcg. of folic acid starting before conception and continuing through the first trimester can significantly reduce the risk of an NTD-affected pregnancy by at least 50 percent.[24,25] The data show that the neural tube defect rate for the state was 0.70 per 1,000 live births (Table 8). Comparisons between the county rates and the state rate indicate that Navajo County had the highest rate for neural tube defects, at 1.22 per 1,000 live births. This was significantly higher than the state rate.

Abdominal Wall Defects

Abdominal wall defects include omphalocele and gastroschisis (Table 9). Gastroschisis is a congenital opening of the abdominal wall, often with protrusion of the intestines. Omphalocele is a membrane-covered protrusion of an abdominal organ through the abdominal wall at the umbilicus. According to a recent study, young mothers are four times as likely as women in their late twenties to have a child with gastroschisis.[26] Other risk factors for gastroschisis are maternal use of cocaine, aspirin, or amphetamines; exposure to solvents; and maternal dietary inadequacy.[27] Table 9 presents the incidence rate for the state at 0.51 per 1,000 live births. Mohave County had the highest incidence rate for abdominal wall defects, at 1.03 per 1,000 live births. There are no statistical differences between the county rates and the state rate for abdominal wall defects.

Heart Defects

This category includes truncus arteriosus, transposition of the great vessels, Tetralogy of Fallot, single ventricle, aortic stenosis, hypoplastic left heart, and total anomalous pulmonary venous return (Table 10). Table 10 shows that the heart defect rate for Gila County was higher than the state rate for heart defects (2.14 vs. 1.49 per 1,000 live births). Navajo County had the second highest heart defect rate, at 2.03 per 1,000 live births. The county rates for heart defects were not found to be statistically different from the state rate.

Table 6
Chromosomal Defects - Incidence Rates by County, 1986-1997
(Live Born Cases per 1,000 Live Births)

COUNTY	CASES 1986-1997	RATE	99% CONFIDENCE INTERVAL
Arizona	1121	1.36	1.26-1.47
Apache	34	1.75	1.07-2.68
Cochise	30	1.49	0.88-2.35
Coconino	32	1.41	0.85-2.20
Gila	15	1.89	0.86-3.55
Graham	9	-	-
Greenlee	3	-	-
La Paz	3	-	-
Maricopa	647	1.32	1.19-1.46
Mohave	24	1.30	0.72-2.16
Navajo	39	1.76	1.12-2.63
Pima	176	1.31	1.07-1.58
Pinal	31	1.24	0.74-1.93
Santa Cruz	18	2.04	1.01-3.64
Yavapai	20	1.26	0.65-2.18
Yuma	40	1.29	0.83-1.92

Chromosomal defects include three-digit codes R01, R02, R03 (see Tables 1-A and B).
- = Insufficient cases for rate and confidence interval calculations.

Table 7
Oral Clefts - Incidence Rates by County, 1986-1997
(Live Born Cases Per 1,000 Live Births)

COUNTY	CASES 1986-1997	RATE	99% CONFIDENCE INTERVAL
Arizona	1467	1.78	1.66-1.90
Apache	59	3.03	2.11-4.21
Cochise	39	1.94	1.23-2.90
Coconino	50	2.21	1.49-3.15
Gila	22	2.77	1.48-4.69
Graham	16	3.06	1.44-5.64
Greenlee	2	-	-
La Paz	3	-	-
Maricopa	796	1.62	1.48-1.78
Mohave	27	1.47	0.84-2.36
Navajo	62	2.80	1.97-3.85
Pima	238	1.77	1.49-2.09
Pinal	52	2.07	1.41-2.94
Santa Cruz	17	1.93	0.93-3.49
Yavapai	32	2.01	1.21-3.12
Yuma	52	1.68	1.14-2.38

Oral Clefts include three-digit codes F01 & F02 (see Tables 1-A and B).
- = Insufficient cases for rate and confidence interval calculations.

Table 8
Neural Tube Defects - Incidence Rates by County, 1986-1997
(Live Born Cases Per 1,000 Live Births)

COUNTY	CASES 1986-1997	RATE	99% CONFIDENCE INTERVAL
Arizona	578	0.70	0.63-0.78
Apache	11	0.57	0.22-1.17
Cochise	9	-	-
Coconino	8	-	-
Gila	9	-	-
Graham	4	-	-
Greenlee	0	-	-
La Paz	3	-	-
Maricopa	358	0.73	0.63-0.84
Mohave	12	0.65	0.27-1.31
Navajo	27	1.22	0.70-1.96
Pima	79	0.59	0.43-0.78
Pinal	15	0.60	0.27-1.13
Santa Cruz	10	1.13	0.42-2.43
Yavapai	11	0.69	0.27-1.43
Yuma	22	0.71	0.38-1.20

Neural Tube defects include three-digit codes A01, A02, A03 & A13 (see Tables 1-A and B).
- = Insufficient cases for rate and confidence interval calculations.

Table 9
Abdominal Wall Defects - Incidence Rates by County, 1986-1997
(Live Born Cases Per 1,000 Live Births)

COUNTY	CASES 1986-1997	RATE	99% CONFIDENCE INTERVAL
Arizona	417	0.51	0.44-0.58
Apache	4	-	-
Cochise	6	-	-
Coconino	12	0.53	0.19-1.12
Gila	6	-	-
Graham	3	-	-
Greenlee	0	-	-
La Paz	0	-	-
Maricopa	236	0.48	0.40-0.56
Mohave	19	1.03	0.46-1.91
Navajo	9	-	-
Pima	82	0.61	0.44-0.78
Pinal	14	0.56	0.18-1.03
Santa Cruz	4	-	-
Yavapai	9	-	-
Yuma	13	0.42	0.17-0.91

Abdominal Wall defects include three-digit codes N02 & N04 (see Tables 1-A and B).
- = Insufficient cases for rate and confidence interval calculations.

Table 10
Heart Defects - Incidence Rates by County, 1986-1997
(Live Born Cases Per 1,000 Live Births)

COUNTY	CASES 1986-1997	RATE	99% CONFIDENCE INTERVAL
Arizona	1229	1.49	1.38-1.60
Apache	20	1.03	0.53-1.78
Cochise	29	1.44	0.84-2.29
Coconino	36	1.59	0.99-2.41
Gila	17	2.14	1.03-3.88
Graham	8	-	-
Greenlee	2	-	-
La Paz	1	-	-
Maricopa	740	1.51	1.37-1.65
Mohave	21	1.14	0.60-1.95
Navajo	45	2.03	1.33-2.95
Pima	199	1.48	1.21-1.75
Pinal	35	1.40	0.86-2.13
Santa Cruz	8	-	-
Yavapai	26	1.63	0.92-2.65
Yuma	42	1.36	0.88-2.00

Heart defects include three-digit codes D01, D02, D03, D04, D51, D52 & D53.
(See Tables 1-A and B.)
- = Insufficient cases for rate and confidence interval calculations.

APPENDIX 1
Conditions Included in the Figures

A general listing of all conditions used to establish the rates shown in the figures in this report is shown below. Some specific inclusions and exclusions are not listed. ABDMP collects data on 140 conditions or variations of conditions. The conditions listed below include over 99% of all cases reported through the ABDMP.

BPA 3-Digit Code*	General Code Descriptor
740 - 759	Congenital anomalies including, but not limited to:
740	Anencephaly and similar anomalies
741	Spina bifida
742	Other anomalies of the nervous system
743	Anomalies of the eye
744	Anomalies of the ear, face, and neck
745	Certain anomalies of the heart
746	Other anomalies of the heart
747	Anomalies of the circulatory system
748	Anomalies of the respiratory system
749	Cleft palate and cleft lip
750	Other anomalies of the upper alimentary tract
751	Anomalies of the digestive system
752	Anomalies of the genital organs
753	Anomalies of the urinary system
754	Certain musculoskeletal deformities
755	Other anomalies of limbs
756	Other musculoskeletal anomalies
757	Congenital anomalies of the integument
758	Chromosomal anomalies
759	Other and unspecified anomalies
ICD-9-CM Code**	
658.80-658.83	Amniotic bands
760.71	Fetal alcohol syndrome

* British Pediatric Association Classification of Diseases
** International Classification of Disease - 9th Edition, Clinical Modification

APPENDIX 2
Conditions (Composite Categories) Shown in the Tables

A listing of the conditions analyzed in the tables contained in this report is shown below.

The 44 conditions listed here can be described almost completely by codes created by the Centers for Disease Control's Metropolitan Atlanta Congenital Defects Program (MACDP). These codes are listed on the left below, with exceptions noted. On the right below are the corresponding British Pediatric Association (BPA) Classification of Diseases codes.

In the tables, a case is listed only once in each MACDP code category, even when it has more than one diagnosis within the category.

MACDP	Condition	BPA Codes		
CENTRAL NERVOUS SYSTEM				
A01	Anencephaly	740.00	740.02	740.03
		740.08	740.10	740.20
		740.21	740.29	
A02	Spina bifida with hydrocephaly	741.00	741.01	741.02
		741.03	741.04	741.05
		741.06	741.07	741.08
		741.09	741.085	741.086
		741.087		
A03	Spina bifida without hydrocephaly	741.90	741.91	741.92
		741.93	741.94	741.98
		741.985	741.99	
A13	Encephalocele	742.00	742.08	742.09
		742.085	742.086	
A15	Hydrocephaly	742.30	742.31	742.38
		742.39		
A16	Microcephaly	742.10		

EYE AND EAR				
B03	Glaucoma	743.20	743.21	743.22
B04	Cataract	743.32	743.325	743.326
B51*	Anophthalmia	743.00		
B52*	Microphthalmia	743.10		
B54*	Ear anomaly with hearing loss	744.00	744.01	744.02
		744.03	744.09	744.21
CARDIAC				
D01	Truncus arteriosus	745.00	745.01	
D02	Transposition of great vessels	745.10	745.11	745.12
		745.18	745.19	
D03	Tetralogy of Fallot	745.20	745.21	746.84
D04	Single ventricle	745.30		
D51*	Aortic stenosis	746.30	746.31	
D52*	Hypoplastic left heart	746.70		
D53*	Total anomalous pulmonary venous	747.42		
RESPIRATORY				
E01	Choanal atresia	748.00		
E06	Agenesis of lung	748.50	748.51	
OROFACIAL - GASTROINTESTINAL				
F01	Cleft palate	749.00	749.01	749.02
		749.03	749.04	749.05
		749.06	749.07	749.09
F02	Cleft lip with or without cleft palate	749.10	749.11	749.12
		749.19	749.20	749.21
		749.22	749.29	
F08	Pyloric stenosis	750.51		
F09	Tracheoesophageal fistula /stenosis	750.30	750.31	750.32
		750.325	750.33	
F14	Stenosis or atresia of duodenum	751.10		
F15	Other stenosis/atresia of small intestine	751.11	751.12	751.19

F16	Stenosis or atresia of rectum or anus	751.21	751.22	751.23
		751.24		
F17	Hirschsprung's Disease	751.30	751.31	751.32
		751.33		
F18	Malrotation of intestine	751.40	751.41	751.42
		751.49	751.495	
F21	Biliary atresia	751.65		
GENITO-URINARY				
H01	Renal agenesis	753.00	753.01	
H06	Obstruction of kidney or ureter	753.20	753.21	753.22
		753.29	753.40	753.42
H09	Bladder or urethra obstruction	753.600	753.61	753.62
		753.63		
MUSCULOSKELETAL				
J03	Dislocation of hip	754.30		
J51*	Complete absence of upper/lower limb	755.20	755.30	755.40
J52*	Phocomelia of limb	755.21	755.31	755.41
K05	Amniotic bands	658.80		
N01	Diaphragmatic hernia	756.61	756.615	756.616
		756.617		
N02	Omphalocele	756.70		
N04	Gastroschisis	756.71		
SYNDROMES				
R01	Down Syndrome (Trisomy 21)	758.00	758.01	758.02
		758.03	758.04	758.09
R02	Patau Syndrome (Trisomy 13)	758.10	758.11	758.12
		758.13	758.19	
R03	Edwards Syndrome (Trisomy 18)	758.20	758.21	758.22
		758.23	758.29	758.295
S02	Fetal alcohol syndrome	760.71	760.718	

* Codes created by California BDMP

APPENDIX 3
Precision of Diagnosis Codes

Often health care professionals qualify a diagnosis, using words to express their level of confidence that the particular diagnosis explains what has been observed when examining, testing or performing a procedure on a patient. If a professional makes a diagnosis using a qualifying term, the ABDMP assigns that diagnosis a "precision code," based on the table below. Higher code numbers indicate higher "levels of precision." Generally, if a diagnosis is made several times with different levels of precision, the diagnosis is assigned the precision code consistent with the most certain diagnosis.

Precision Code/Qualifying Terms

1	Not stated. (For mental retardation and cerebral palsy diagnoses ONLY)
2	"Probably not," "ruled out," "NO"
3	"vs" (versus) or "or"
4	"Rule out" included in diagnosis (i.e., rule out anencephaly), "doubtful," "equivocal", "questionable," "r/o," "borderline," "somewhat," "mildly"
5	"Suggestive of"
6	"Suspected," "suspicious"
7	"Possible," "may have," "could be," "felt to be," "perhaps," "consider"
8	"Consistent with," "most likely"
9	"Compatible with," "like," "appears"
10	"Probable," "presume"
11	(This code is not currently used.)
12	Precise diagnosis, "characteristic of"
13	Precise diagnosis with congestive heart failure or medicated with Digoxin, Drisdol, Chlorothiazide, Lasix, Lanoxin, Aldactone or other diuretics (only for VSD, PDA, ASD, or patent foramen ovale)

APPENDIX 4
Abbreviations

ABDMP	- Arizona Birth Defects Monitoring Program
ADHS	- Arizona Department of Health Services
BPA	- British Pediatric Association
CBDMP	- California Birth Defects Monitoring Program
CDC	- Centers for Disease Control and Prevention
CRS	- Children's Rehabilitative Services (ADHS)
ICD	- International Classification of Disease
MACDP	- Metropolitan Atlanta Congenital Defects Program

APPENDIX 5
Exclusion List - ABDMP
Non-reportable Birth Defects Cases

The following potential cases are not included in the ABDMP report for 1994 and 1997:

- Duplicate abstracts and/or duplicated anomalies (cases with multiple abstracts; child seen at more than one facility), i.e., duplicate cases are merged and counted once.

- "Possibles" abstracted for review and consideration and subsequently determined to have conditions or defects that were not reportable according to the CDC and CBDMP lists of "excludable conditions."

- Babies born to mothers whose residences are out-of-state or out-of-country (i.e., nonresident cases).

- "Negatives," that is, conditions that were ruled-out during case-finding and medical record review.

- "No Match"cases. A birth certificate was not on file and the state of birth could not be confirmed as Arizona.

- Cases among aborted fetuses less than 20 weeks gestation and weighing less than 500 grams. These cases were excluded because there was no reliable denominator that could be used to generate a birth defect rate.

- Prenatally diagnosed cases that did not result in a known live birth or stillbirth are not included. The ABDMP is not currently visiting prenatal diagnostic centers to identify cases.

- Defects with a "precision of diagnosis" code 1-7 are excluded. Only those defects diagnosed at the higher levels of precision (8 or above) are included. Refer to Appendix 3 for list of precision of diagnosis codes.

- Cases only diagnosed outside of the hospital setting are not included in the ABDMP. The Arizona Children's Rehabilitative Services Clinics are the only outpatient facilities we ascertain cases from.

APPENDIX 6
Definitions

Race and Ethnicity: .

Arizona birth, death, and fetal death certificates contain information on both race and ethnicity, allowing for the simultaneous classification/differentiation of persons classified as “Whites,” “Blacks,” “Native American,” and “Asian” and as of Hispanic origin or non-Hispanic origin. The Hispanic category consists of mothers who answered “White” to race and “Hispanic” to the Hispanic origin question. In this report, the race and ethnic classification is as follows:

• “White” refers to White non-Hispanic

• “Hispanic” refers to White Hispanics

• “Black” refers to African Americans

• “Native American” includes all Native American tribes, as well as Aleut and Eskimo

• “Other” includes persons who are Asian, unclassified, or did not provide a response to race question on the certificate.

Native American counts refer to all Native Americans living on and off the reservations.

Since the Arizona Birth Defects Monitoring Program does not collect the race and ethnicity of the child nor its parents, race and ethnicity of the child and its parents are derived from the classification used by Vital Statistics, upon the merging of the ABDMP data records with the Birth, Death, and Fetal Death Certificate data.

APPENDIX 7
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APPENDIX 8
Birth Weight

The Arizona Birth Defects Monitoring Program monitors the distribution of birth weight. The data is obtainable from the birth certificate and may allow the detection of major shifts over time in the proportion of newborns with low birth weight. [27,28] Of note, the proportion of babies > 4,000 grams at birth appears to be decreasing, perhaps indicating improved control of gestational diabetes.